Pediatric Manual

Appendix 1: Pediatric Data Collection System

Effective with Cases diagnosed 1/1/2024 and Forward

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Version 1.1

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Contents

Pediatric Data Collection System

The Pediatric Data Collection System (DCS) has been developed to collect Pediatric staging and site-specific data item (SSDI) information. The staging elements collected are based on the *Toronto Childhood Cancer Staging Guidelines, Version 2, along* with additional data items for surveillance purposes. The pediatric data collection system also allows for expansion to develop further staging information that is not covered in the Toronto Guidelines.

This new data collection system is patterned after SEER's Extent of Disease data collection system, which has been in use since the 1970's and had a major update in 2018, and the SSDI manual, which was developed in 2018. This data collection system will allow the following:

- Permitting staging of the most comprehensive set of patients
- Reporting and monitoring trends in cancer incidence and outcomes
- Supporting and promoting research for pediatric cancers
- Enabling and ensuring ongoing continuity of staging trends over time reflecting the combination of clinical and pathologic information

The Pediatric DCS applies to ages 00-39 (for some histologies all ages) and specific primary site/histology combinations (see 9617: Pediatric ID for a complete listing). This covers Pediatric and Adolescence/Young Adult (AYA) patients. Additional primary site/histology combinations may be added in the future.

The Pediatric DCS uses all information available in the medical record; in other words, it is a combination of the most precise clinical and pathological documentation of the extent of disease.

There are 4 main data items in the Pediatric DCS, each of which is discussed in detail.

1. Pediatric Primary Tumor
2. Pediatric Regional Nodes
3. Pediatric Mets
4. Applicable SSDIs (Schema-dependent)

This manual is effective for all cases diagnosed 1/1/2024 and after.

Send questions, suggestions, and corrections to:

Ask a SEER Registrar

Choose subject: Pediatric

Introduction to the Toronto Childhood Cancer Staging System

Staging for specific pediatric cases will be implemented in the US for the first time in 2024, based on the *Toronto Childhood Cancer Staging Guidelines, Version 2*. Below is a brief introduction of what are the Toronto Childhood Cancer Staging Guidelines are and how they are used.

"A consensus meeting was convened in 2014 by the Union for International Cancer Control (UICC), the Dana-Farber Cancer Institute and the Hospital for Sick Children, Toronto to address the lack of consistent information on childhood cancer stage in population registries.[1] For each of a subset of the major childhood cancer diagnostic groups/subgroups, the meeting reviewed all disease-specific cancer staging systems currently in use and recommended the one most suitable for use by population-based cancer registries. The recommended staging systems are listed as the Toronto Paediatric Cancer Stage Guidelines.

The Guidelines recommended disease-specific staging systems for Acute Lymphoblastic Leukemia, Hodgkin lymphoma, Non-Hodgkin lymphoma, Ependymomas, Astrocytoma, Medulloblastoma, Neuroblastoma, Retinoblastoma, Renal Tumors, Hepatoblastoma, Malignant Bone Tumors, Rhabdomyosarcoma, Non-Rhabdomyosarcoma soft tissue sarcomas, Germ Cell Tumors (Ovary and Testicular).

The Guidelines were successfully tested in practice for their feasibility and validity.[3] They are endorsed by the UICC TNM Prognostic Factors project, the European Network of Cancer Registries (ENCR), the Group for Cancer Epidemiology and Registration in Latin Language Countries (GRELL) and the African Network of Cancer Registries (ANCR) and published in the UICC TNM Classification of Malignant Tumours 8th Edition." [1, 2, 3]

1. Aitken JF, Youlden D, O'Neill L, Gupta S, Frazier AL, eds. Childhood cancer staging for population registries according to the Toronto Childhood Cancer Stage Guidelines – Version 2. Cancer Council Queensland and Cancer Australia: Brisbane, Australia; 2021.
2. Gupta S, Aitken JF, Bartels U, Brierley J, Dolendo M, Friedrich P, Fuentes-Alabi S, Garrido CP, Gatta G, Gospodarowicz M, Gross T, Howard SC, et al. Paediatric cancer stage in population-based cancer registries: the Toronto consensus principles and guidelines. *The Lancet Oncology* 2016;**17**: e163-72.
3. Gupta S, Aitken J, Bartels U, Bhakta N, Bucurenci M, Brierley JD, De Camargo B, Chokunonga E, Clymer J, Coza D, Fraser C, Fuentes-Alabi S, et al. Development of paediatric non-stage prognosticator guidelines for population-based cancer registries and updates to the 2014 Toronto Paediatric Cancer Stage Guidelines. Lancet Oncol 2020;21: e444-e51.

The Toronto Childhood Cancer Stage Guidelines can be found at http://www.iacr.com.fr/8index.php?option=com_content&view=article&id=153&Itemid=657.

In 2020, the US National Cancer Institute (NCI) launched the Childhood Cancer Data Initiative (CCDI). A key component of this initiative is the development and maintenance of the National Childhood Cancer Registry (NCCR), a public health surveillance data resource whose primary goal is to gather data from every child, adolescent, and young adult diagnosed with a childhood cancer ultimately to better understand the causes, outcomes, effective treatments, and later effects of cancer among children, adolescents, and young adults in the US. Part of the NCCR project included adopting the Toronto Staging Guidelines and implementing new data items to get the Toronto Stage and other relevant clinical factors. Data collected will be comparable with international data and provide information to assess the burden of pediatric cancers worldwide.

General Coding Instructions for Pediatric Data Items

The Pediatric Data Collection System has three data items: Pediatric Primary Tumor, Pediatric Regional Nodes, and Pediatric Mets. This data collection system is new for the cancer registry field in the US and Canada and applies to individuals mostly between the ages of 00-39 and is applicable for select primary site/histology combinations for 2024.

- Every combination of primary site and histology will be accounted for in a Pediatric Schema. Those primary site/histology combinations not defined by the *Toronto Childhood Cancer Staging Guidelines, Version 2*, will be grouped into an "other" schema, which will not require any additional input from the registrar.

Do not use this system for any cases diagnosed prior to 1/1/2024.

***Note*: ALWAYS check site-specific Pediatric 2024 schemas for exceptions and/or additional information.**

General Guidelines

1. Pediatric schemas apply to ages 00-39 and specific primary site/histology combinations. Many of the pediatric schemas are based primarily on histology.
    a. There are some histologies that will apply to all ages (e.g., Neuroblastoma, Retinoblastoma).
    b. The software will determine which cases will go into a specific Pediatric Schema.

2. For ALL sites, the Pediatric DCS is based on a combined clinical and operative/pathological assessment. Gross observations at surgery are particularly important when all malignant tissue cannot be or was not removed.
    a. In the event of a discrepancy between pathology and operative reports concerning excised tissue, priority is given to the pathology report.

3. Pediatric DCS should include all information available within **four months of diagnosis** in the absence of disease progression or upon completion **of surgery(ies)** in the first course of treatment, whichever is longer.

4. Information for Pediatric DCS from a surgical resection **after neoadjuvant treatment may be used**, but **ONLY** if the extent of disease is greater than the pre-treatment clinical findings.
    a. Exception: For the schemas where Pediatric Primary tumor is based on surgical resection only, findings from a surgical resection post-neoadjuvant therapy can be used.

5. Disease progression, including metastatic involvement, known to have developed after the initial stage workup, should be excluded when coding the Pediatric fields.

6. Autopsy reports are used in coding Pediatric just as are pathology reports, applying the same rules for inclusion and exclusion.

7. Death Certificate only (DCO) cases
    a. Code the following for DCO's. If a data item has a default value (888 or 88), then code the default value
        i) Pediatric Primary Tumor: 999 (unless default is 888)
        ii) Pediatric Regional Nodes: 999 (unless default is 888)
        iii) Pediatric Mets: 99 (unless default is 88)

8. Pediatric Schema-specific guidelines take precedence over general guidelines. Always read the information pertaining to a specific primary site or histology schema.

Ambiguous Terminology

Most of the time, registrars will find definitive statements of extension/involvement; however, for those situations where extension/involvement is described with non-definitive (ambiguous) terminology, use the guidelines below to interpret and determine the appropriate assignment of Pediatric Primary Tumor, Pediatric Regional Nodes or Pediatric Mets.

Determination of the cancer stage is both a subjective and objective assessment by the physician(s) of how far the cancer has spread. When it is not possible to determine the extent of involvement because terminology is ambiguous, look at the documentation that the physician used to make informed decisions on how the patient is being treated. For example, assign the Pediatric fields based on extension/involvement when the patient was treated as though adjacent organs or nodes were involved.

Use the following lists to interpret the intent of the clinician ONLY when further documentation is not available and/or there is no specific statement of extension/involvement in the medical record. The clinician's definitions/descriptions and choice of therapy have priority over these lists because individual clinicians may use these terms differently.

> ***Note 1:*** Terminology in the schema takes priority over this list. Some schemas interpret certain words as involvement, such as 'encasing' the carotid artery for a head and neck site or "abutment," "encases," or "encasement" for pancreas primaries.
>
> ***Note 2***: Use this list only for EOD 2018, Summary Stage 2018, or the Pediatric Data Collection System
>
> ***Note 3:*** This is not the same list used for determining reportability as published in the SEER manual, Hematopoietic Manual or in Section 1 of the Standards for Oncology Registry Entry (STORE). This is not the same list of ambiguous terminology provided in the Solid Tumors Rules published and maintained by the SEER Program

Use the following lists as a guide ***when no other information is available***

Involved

Adherent	Incipient Invasion
Apparent(ly)	Induration
Appears to	Infringe/infringing
Comparable with	Into*
Compatible with	Intrude
Consistent with	Most likely
Contiguous/continuous with Encroaching upon*	Onto*
Extension to, into, onto, out onto	Overstep
Features of	Presumed
Fixation to a structure other than primary**	Probable
Fixed to another structure**	Protruding into (unless encapsulated)
Impending perforation of	Suspected
Impinging upon	Suspicious
Impose/imposing on	To*
	Up to

Not involved

Abuts	Extension to without invasion/involvement of
Approaching	Kiss/kissing
Approximates	Matted (except for lymph nodes)
Attached	Possible
Cannot be excluded/ruled out	Questionable
Efface/effacing/effacement	Reaching
Encased/encasing	Rule out
Encompass(ed)	Suggests
Entrapped	Very close to
Equivocal	Worrisome

* Interpret as involvement whether the description is clinical or operative/pathologic

** Interpret as involvement of the other organ or tissue

Pediatric Data Items

9617: Pediatric ID

Item Length: 5
NAACCR Item #: 9617
XML NAACCR ID: pediatricId
NAACCR Alternate Name: None
Active years: 2024+

Description

The derived values in this data item link Site-Specific Data Items with the appropriate site/histology grouping and account for the combination of primary site and histologies that are being collected for Pediatric Cancers starting in 2024. The values for this data item are derived based on primary site and histology and are initially based on the Toronto Childhood Cancer Stage Guidelines. The derived values link Site-Specific Data Items with the appropriate site/histology grouping.

- *For example*, the Pediatric ID for a Neuroblastoma is 4a. This value links the Staging and Site-Specific Data Items associated with Neuroblastoma: 9611: Intl Neuroblastoma Risk Grp Stag Sys (INRGSS), 9614: n-MYC Amplification, and 9610: Intl Neuroblastoma Path Prog Class (INPC)

Rationale

The purpose of the Pediatric ID is to link the appropriate Site-Specific Data Items and Pediatric Stage Data Items (Pediatric Primary Tumor, Pediatric Regional Nodes, Pediatric Mets) with the patient's primary site/histology. Each Site-Specific Data Item (SSDI) and definitions for the Pediatric Stage Data Items apply only to selected primary sites, histologies, and years of diagnosis.

Definition

Beginning on January 1, 2024, registries will start collecting SSDIs that are for Pediatric Cancer patients only. The Pediatric IDs are based on the *Toronto Childhood Cancer Staging Guidelines, Version 2*.

Pediatric ID Table

Note: Unless noted otherwise, histology, behavior, and primary site groupings apply to ages 0-39 years of age.

Pediatric ID/Name	Criteria	SSDIs
1a1: Acute Lymphoblastic Leukemia (ALL)	9811/3-9819/3 ,9837/3; *C000-C809*	9621: White Blood Cell Count
2a: Hodgkin Lymphoma	9650/3-9653/3, 9655/3, 9659/3, 9663/3: *C000-C809*	3812: B Symptoms
2b2: NHL: Mature B-cell neoplasms	9731/3, 9732/3, 9734/3: *C000-C809* 9671/3, 9673/3, 9678/3-9680/3, 9688/3-9691/3, 9695/3, 9698/3, 9699/3, 9735/3, 9737/3-9738/3, 9761/3-9762/3, 9765/3-9766/3, 9769/3, 9823/3, 9970/3; *C000-C424, C470-C809*	
2b3: NHL: Mature T-cell and NK-cell neoplasms	9702/3, 9705/3, 9714/3-9717/3, 9724/3, 9767/3-9768/3, 9827/3: *C000-C424, C470-C809*	

Pediatric ID/Name	**Criteria**	**SSDIs**
2b4: NHL: NOS	9591/3: (includes all Schema Discriminators): *C000-C424, C470-C809*	
2c: NHL: Burkitt lymphoma	9687*/3; C000-C424, C470-C809*	
3a: Ependymoma	9383, 9391-9394, 9396: *C710-C729 (All ages)*	
3b: Astrocytomas	9380, 9384, 9400-9411, 9420-9424, 9440-9442, 9445: *C700-C729*	3940: BRAF Mutational Analysis
3c1: Medulloblastoma	9470-9472, 9474-9478, 9480: *C700-C729 (All ages)*	
3c2: Medulloblastoma: pNET	9473: *C700-C729 (All ages)*	
3c3: Medulloblastoma: Medulloepithelioma	9501-9504: *C700-C729 (All ages)*	
3c4: Medulloblastoma: Atypical teratoid/rhabdoid tumors	9508: *C700-C729 (All ages)*	
3e3: Medulloblastoma: Pineoblastoma	9362: *C700-C729, C753 (All ages)*	
4a: Neuroblastoma	9490/3, 9500/3: *C000-C809* *All ages*	9611: Intl Neuroblastoma Risk Grp Stag Sys (INRGSS) 9614: n-MYC Amplification 9610: International Neuroblastoma Pathology Prognostic Classification (INPC)
5: Retinoblastoma	9510/3-9514/3: *C690-C699 (All ages)*	9627: IRSS Stage for Eye-2
6a1: Renal Tumors: Nephroblastoma	8959/3, 8960/3: *C649, C659 (All ages)*	3801: Chromosome 1p: Loss of Heterozygosity 9600: Chromosome 16q: Loss of Heterozygosity 9601: Chromosome 1q Status 9608: EWSR1-FLI1 fusion (6a4 only)
6a2: Renal Tumors: Rhabdoid Renal Tumor	8963/3: *C649 (All ages)*	3801: Chromosome 1p: Loss of Heterozygosity 9600: Chromosome 16q: Loss of Heterozygosity 9601: Chromosome 1q Status 9608: EWSR1-FLI1 fusion (6a4 only)
6a3: Renal Tumors: Kidney Sarcomas	8964/3-8967/3: *C649, C659*	3801: Chromosome 1p: Loss of Heterozygosity 9600: Chromosome 16q: Loss of Heterozygosity 9601: Chromosome 1q Status 9608: EWSR1-FLI1 fusion (6a4 only)

Pediatric ID/Name	Criteria	SSDIs
6a4: Renal Tumors: Ewing Sarcoma of Kidney	9364/3: *C649 (All ages)*	3801: Chromosome 1p: Loss of Heterozygosity 9600: Chromosome 16q: Loss of Heterozygosity 9601: Chromosome 1q Status 9608: EWSR1-FLI1 fusion (6a4 only)
6c: Renal Tumors: Unspecified Malignant Renal Tumors	8000/3-8005/3: *C649*	3801: Chromosome 1p: Loss of Heterozygosity 9600: Chromosome 16q: Loss of Heterozygosity 9601: Chromosome 1q Status 9608: EWSR1-FLI1 fusion (6a4 only)
7a: Hepatoblastoma	8970/3: *C220 (All ages)*	9626: Pretext Clinical Staging
8a: Malignant Bone Tumors: Osteosarcoma	9180/3-9187/3, 9191/3-9195/3, 9200/3: *C400-C419, C760-C768, C809*	
8b: Malignant Bone Tumors: Chondrosarcoma	9210/3, 9220/3-9221/3, 9230/3, 9240/3-9243/3: *C400-C419, C760-C768, C809*	
8c: Malignant Bone Tumors: Ewing	9260/3: *C400-C419, C760-C768, C809* 9363/3-9365/3: *C400-C419*	9608: EWSR1-FLI1 fusion
8d: Malignant Bone Tumors: Other specified	8810/3-8812/3, 8823/3, 8830/3, 9250/3, 9261/3-9262/3, 9270/3-9275/3, 9280/3-9282/3, 9290/3, 9300/3-9302/3, 9310/3-9312/3, 9320/3-9322/3, 9330/3, 9340/3-9342/3, 9370/3-9372/3: *C400-C419*	
8e: Malignant Bone Tumors: Unspecified	8000/3-8005/3, 8800/3, 8801/3, 8803/3-8805/3: *C400-C419*	
9a: Rhabdomyosarcoma	8900/3-8905/3, 8920/3, 8991/3: *C000-C809* 8910/3, 8912/3: *C000-C809 (All ages)*	9609: FOXO1 Gene Rearrangements
9b: Non-Rhabdo-myosarcoma: Fibrosarcomas	8810/3, 8811/3, 8813/3–8815/3, 8821/3, 8823/3, 8834/3–8835/3: *C000-C399, C440-C768, C809* 8820/3, 8822/3, 8824/3-8827/3, 9150/3, 9160/3, 9491/3, 9540/3-9571/3, 9580/3: *C000-C809*	

Pediatric ID/Name	Criteria	SSDIs
9d: Non-Rhabdo-myosarcoma: Other specified	8587/3, 8710/3–8713/3, 8806/3, 8831/3–8833/3, 8836/3, 8840/3–8842/3, 8850/3–8858/3, 8860–8862, 8870, 8880, 8881, 8890–8898, 8921, 8982, 8990, 9040–9044, 9120–9125, 9130–9133, 9135, 9136, 9141, 9142, 9161, 9170–9175, 9231, 9251, 9252, 9373, 9581: *C000-C809* 8830/3: *C000-C399, C440-C768, C809* 8963/3: *C000-C639, C659-C699, C739-C768, C809* 9180/3, 9210/3, 9220/3, 9240/3: *C490-C499* 9260/3: *C000-C399, C470-C759* 9364/3: *C000-C399, C470-C639, C659-C699, C739-C768, C809* 9365/3: *C000-C399, C470-C639, C659-C768, C809*	
9e: Non-Rhabdomyosarcoma: Unspecified	8800/3-8805/3: *C000-C399, C440-C768*	
10c1: Testicular	9060/3-9065/3, 9070/3-9073/3, 9080/3-9085/3, 9090/3-9091/3, 9100/3-9101/3: *C620-C629*	3923: S Category Clinical 3924: S Category Pathological
10c2: Ovarian	9060/3-9065/3, 9070/3-9073/3, 9080/3-9085/3, 9090/3-9091/3, 9100/3-9101/3: *C569*	
99999: Adult/non-pediatric	Remaining primary site/histology combinations	

9618: Pediatric ID Version Current

Item Length: 5
NAACCR Item #: 9618
XML NAACCR ID: pediatricIdVersionCurrent
NAACCR Alternate Name: None
Active years: 2024+

Description
This item indicates the version of Pediatric component of the SEER Staging API used to assign the 2024 and later pediatric staging fields of Pediatric ID and Pediatric input fields. This data item is recorded the first time the Pediatric ID is determined and should be updated each time the related input fields are modified.

Rationale
Over time, the definitions for the Pediatric ID and the input codes and instructions for other related Pediatric data items may change. This item identifies the correct interpretation of information recorded.

Codes

Code	Description
0.3	BETA: 10/17/2022 – Kentucky Cancer Registry Only
0.4	BETA: 11/2/2022 – Kentucky Cancer Registry Only
0.5	BETA: 5/5/2023 – Kentucky Cancer Registry Only
1.0	10/16/2023 release

Codes
Pediatric ID Version Current is a code with up to 2 digits, a decimal and then up to 2 more digits. (e.g., 1.5, 10.12). The first two digits represent the major version number; the second two digits represent minor version changes. The minimum allowable value would be “0.3”. The maximum allowable value would be “99.99”. Blanks would not be allowed.

This data item will be generated by registry software. No coding instructions are required.

9619: Pediatric ID Version Original

Item Length: 5
NAACCR Item #: 9619
XML NAACCR ID: pediatricIdVersionOriginal
NAACCR Alternate Name: None
Active years: 2024+

Description
This item indicates the version of Pediatric component of the SEER Staging API used to assign the 2024 and later pediatric staging fields of Pediatric ID and Pediatric input fields. This data item is recorded the first time the Pediatric ID is determined. This data item should not be updated each time the related input fields are modified.

Rationale
Over time, the definitions for the Pediatric ID and the input codes and instructions for other related Pediatric data items may change. This item identifies the correct interpretation of information recorded.

Codes

Code	Description
0.3	BETA: 10/17/2022 – Kentucky Cancer Registry Only
0.4	BETA: 11/2/2022 – Kentucky Cancer Registry Only
0.5	BETA: 5/5/2023 – Kentucky Cancer Registry Only
1.0	10/16/2023 release

Pediatric ID Version Current is a code with up to 5 digits, a decimal and then up to 2 more digits. (e.g., 1.5, 10.12). The first two digits represent the major version number related to diagnosis year; the second two digits represent minor version changes with the diagnosis years. The minimum allowable value would be “0.5”. The maximum allowable value would be “99.99”. Blanks would not be allowed.

This data item will be generated by registry software. No coding instructions are required.

9620: Toronto Version Number

Item Length: 1
NAACCR Item #: 9620
XML NAACCR ID: torontoVersionNumber
NAACCR Alternate Name: None
Active years: 2024+

Description
This item indicates the Toronto Staging Version number that the Pediatric staging is based on. This data item is recorded based on when the case is abstracted.

Rationale
The Pediatric Staging API is based on the Toronto Staging System, so named because the definitions were first discussed at a meeting held in Toronto. Over time, the definitions will be revised, and the Pediatric staging API will be updated accordingly. This value captures the Toronto version which the current Pediatric Staging API is based on.

Codes

Code	Description
1	First version of Toronto Staging (Pediatric Staging versions 0.3-0.5)
2	Second version of Toronto Staging (Pediatric Staging version 1.0 and later)

This data item will be generated by registry software. No coding instructions are required.

9623: Pediatric Primary Tumor

Item Length: 3
NAACCR Item #: 9623
XML Parent-NAACCR ID: pediatricPrimaryTumor
Active years: 2024+

Description

Pediatric Primary Tumor is part of the Pediatric Staging data collection system and is used to classify contiguous growth (extension) of the primary tumor within the organ of origin or its direct extension into neighboring organs. See also Pediatric Regional Nodes **[NAACCR Data item #9624],** and Pediatric Mets **[NAACCR Data item #9625]**. Effective for cases diagnosed 1/1/2024 and forward.

Rationale

Pediatric Primary Tumor is used to calculate Derived Pediatric T (when applicable) **[NAACCR Data item #9607].** Derivation will occur at the level of the central registry.

See the most current version of SEER*RSA (Pediatric tab) for rules and site-specific codes and coding structures.

Code	Description
000	In situ, intraepithelial, noninvasive, non-infiltrating
	SCHEMA-SPECIFIC CODES WHERE NEEDED
800	No evidence of primary tumor
888	Not applicable
999	Unknown; extension not stated Primary tumor cannot be assessed Not documented in patient record Death Certificate Only

Coding Instructions

1. **Assign the farthest documented contiguous involvement of the primary tumor**. Code the farthest documented contiguous direct extension/involvement of tumor away from the primary site. If an involved organ or tissue is not specifically mentioned in the code descriptions, approximate the location from listed structures in the same anatomic area and assign the appropriate code based on that information. Pediatric Primary Tumor codes are hierarchical except for code 800.
2. **Pathological codes**. Some schemas have Pediatric extension/involvement codes that can only be used when there is a surgical resection.
3. **Pathological findings take priority over clinical findings.**
    a. Assign the highest code representing the greatest extension/involvement pathologically (based on pathology report), when available
    b. If there is no applicable pathology, assign the highest code representing the greatest extension/involvement clinically. Imaging takes precedence over physical examination
    c. If extension/involvement is positive based on imaging and/or physical exam, but is confirmed to be negative on pathological exam, then code Pediatric Primary Tumor based on the pathological findings

4. **Neoadjuvant (preoperative) therapy**: If the patient receives neoadjuvant (preoperative) systemic therapy (chemotherapy, immunotherapy) or radiation therapy, code the clinical information if that is the farthest extension/involvement documented. If the post-neoadjuvant surgery shows more extensive disease, code the extension/involvement based on the post-neoadjuvant information. If the clinical and pathological information are the same, code the extension/involvement based on the clinical information.

5. When multiple tumors are reported as a single primary, code the furthest direct extension/involvement from any tumor.

6. **Code 800** when there is no evidence of the primary tumor (occult primary).
    a. Use code 800 when clinically and/or pathologically there is no evidence of the primary tumor. This code does **not** apply to those cases where a biopsy removes all the tumor and there is no residual tumor on the surgical resection

7. When Pediatric Primary Tumor is coded 800, code Tumor Size to 000.

8. **Code 999**
    a. Assign code 999 when there is no information on primary tumor extent

    b. Code 999 is to be used by default for death certificate only (DCO) cases

9. **Document choice of Pediatric Primary Tumor code in text.** It is strongly recommended that the assessment of the primary tumor extension/involvement be documented, as well as the choice of the Pediatric Primary Tumor code in a related STAGE text field on the abstract. While primary tumor extension/involvement can be found in a variety of places, it's most found in a pathology and/or operative report.

9624: Pediatric Regional Nodes

Item Length: 3
NAACCR Item #: 9624
XML Parent-NAACCR ID: pediatricRegionalNodes
Active years: 2024+

Description

Pediatric Regional Nodes is part of the Pediatric Staging data collection system and is used to classify the regional lymph nodes involved with cancer at the time of diagnosis**.** See also Pediatric Primary Tumor **[NAACCR Data item #9623]** and Pediatric Mets **[NAACCR Data item #9625]**. Effective for cases diagnosed 1/1/2024 and forward.

Rationale

Pediatric Regional Nodes is used to calculate Derived Pediatric N (when applicable) **[NAACCR Data item #9605].** Derivation will occur at the level of the central registry.

See the most current version of SEER*RSA (Pediatric tab) for rules and site-specific codes and coding structures.

Code	Description
000	No regional lymph node involvement
	SCHEMA-SPECIFIC CODES WHERE NEEDED
800	Regional lymph node(s), NOS Lymph node(s), NOS
888	Not applicable
999	Unknown; regional lymph node(s) not stated Regional lymph node(s) cannot be assessed Not documented in patient record Death Certificate Only

Coding Instructions

1. **Record the specific involved regional lymph node chain(s) farthest from the primary site.** Regional lymph nodes are listed for each schema. Pediatric Regional Nodes are hierarchical, except for code 800.

    a. Generally, the regional lymph nodes in the chain(s) closest to the primary site have lower codes, while nodes farther away from the primary or in farther lymph node chains have higher codes, although there are exceptions due to lymph drainage patterns.

    b. If a lymph node chain is not listed, check the abstractor notes in SEER*RSA, Appendix C of the Hematopoietic Manual, an anatomy textbook, ICD-O-3, or a medical dictionary for a synonym. **If the lymph node chain or its synonym are not listed in regional lymph nodes, code the involved node(s) in Pediatric Mets.**

        i. Tip for coding lymph nodes: If not possible to determine if a lymph node is regional or distant, check the scheme for a site that is nearby.

    c. Make sure your Pediatric Lymph Node code agrees with Regional Nodes Positive
        i. If Regional Nodes Positive = 01-90, 95, 97, this indicates that regional lymph nodes are involved, and Pediatric Lymph Nodes should be coded appropriately

2. CLINICAL vs PATHOLOGICAL codes
    a. Some schemas have Pediatric regional node codes that are noted as "clinical assessment only" or "pathological assessment only.
        i. Clinical assessment codes should be used only when there is a clinical work up and there is no surgical resection of the primary tumor or site. This includes physical exam, FNA, needle core biopsy, sentinel node biopsy, or lymph node excision.

            1. *Exception:* If patient has neoadjuvant therapy, and the clinical assessment is greater than the pathological assessment, then the clinical assessment code would take priority

        ii. Pathological assessment codes can be used when there is a surgical resection of the primary tumor or site in conjunction with a FNA, Sentinel Lymph Node biopsy or lymph node dissection. The FNA or sentinel lymph node biopsy can be done during the clinical workup and then followed by a negative lymph node dissection

3. **Pathological findings take priority over clinical findings**: It is not necessary to biopsy every lymph node in the suspicious area to disprove involvement. See next section for coding instructions when neo-adjuvant therapy is administered.

    a. Code the lymph node involvement at diagnosis pathologically (based on pathology report), when available.

    b. If there is no applicable pathology, assign lymph node involvement based on clinical findings. Imaging takes precedence over physical examination.

    c. If nodes are determined positive based on imaging and then confirmed to be negative on pathological exam, then code Pediatric Regional Nodes based on the negative pathological findings.

    ***Exception*:** Assign code 800, "Regional lymph node(s), NOS or Lymph node(s), NOS" only when there is lymph node involvement, but no available information regarding the specific node(s) involved.

4. **Neoadjuvant (preoperative) therapy:** If the patient receives neoadjuvant (preoperative) systemic therapy (chemotherapy, immunotherapy) or radiation therapy, code the clinical information if that is the most extensive lymph node involvement documented. If the post-neoadjuvant surgery shows more extensive lymph node involvement, code the regional nodes based on the post-neoadjuvant information. If the clinical and pathological information are the same, code regional lymph nodes based on the clinical information.

5. **Terms meaning lymph node involvement:** For solid tumors, the terms "fixed" or "matted" and "mass in the hilum, mediastinum, retroperitoneum, and/or mesentery" (with no specific information as to tissue involved) are recorded as involvement of lymph nodes.

    a. Other terms, such as "palpable," "enlarged," "visible swelling," "shotty," or "lymphadenopathy" should be ignored for solid tumors, unless there is a statement of involvement by the clinician, or the patient was treated as though regional nodes were involved.

    ***Example**: Enlarged renal hilar nodes found on CT, positive for cancer. Record as involvement of lymph nodes.*

    b. The terms "homolateral," "ipsilateral," and "same side" are used interchangeably.

6. **Accessible lymph nodes:** For "accessible" lymph nodes that can be observed, palpated, or examined without instruments, such as the regional nodes for the breast, oral cavity, salivary gland, skin, thyroid, and other organs, look for some description of the regional lymph nodes. **A statement such as "remainder of examination negative" is sufficient to code 000 negative regional lymph nodes.**

   ***Note**:* If there is mention of a clinical evaluation but no mention of positive lymph nodes, assign code 000.

7. **Inaccessible lymph nodes:** For certain primary sites, regional lymph nodes are not easily examined by palpation, observation, physical examination, or other clinical methods. These are lymph nodes within body cavities that in most situations cannot be palpated, making them inaccessible. Bladder, colon, corpus uteri, esophagus, kidney, liver, lung, ovary, prostate, and stomach are examples of inaccessible sites (this is not an all-inclusive list). When Pediatric Primary Tumor is low stage/Localized and standard treatment is done, it is sufficient to code 000 for negative regional lymph nodes.

8. Code Pediatric Regional Nodes 000 (negative) instead of 999 (unknown) when **ALL** three of the following conditions are met:
   a. There is no mention of regional lymph node involvement in the physical examination, pre-treatment diagnostic testing, or surgical exploration
   b. The patient has localized disease
   c. The patient receives what would be the standard treatment to the primary site (treatment appropriate to the stage of disease as determined by the physician), or patient is offered usual treatment but refuses it

   These guidelines apply only to localized cancers. Assign code 999 when there is reasonable doubt that the tumor is localized.

   ***Example**:* When there is evidence that a testis cancer has extended to the vas deferens, (regional disease) and regional lymph node involvement is not mentioned, it would be correct to code 999 for unknown lymph node involvement in the absence of any specific information regarding regional nodes.

9. **Direct tumor extension/involvement into lymph node:** If direct extension/involvement of the primary tumor into a regional lymph node is shown, code the involved node(s) in Pediatric Regional Nodes.

10. **Code 800.** Use code 800 for the following situations:
    a. Lymph node assignment for the Pediatric schema is based on location (specifically listed lymph nodes) and the only documentation available is that lymph nodes are involved
    b. Lymph node assignment for the Pediatric is based on number and/or size and the only documentation available is that lymph nodes are involved
    c. Statement of "regional lymph nodes involved," with no further information on location, number and/or size.
    d. Unidentified nodes included with the resected primary site
       i. Nodes may be identified in the operative or pathology report (including the final diagnosis), microscopic or gross description
    e. Lymph nodes which are not specified as regional or distant should be assumed to be regional nodes

11. **Code 999**

a. Assign code 999 when there is no information on regional lymph node involvement and the primary tumor is not localized

b. Code 999 is to be used by default for death certificate only (DCO) cases

12. **Document choice of Pediatric Regional Nodes code in text.** It is strongly recommended that the positive and negative assessment of regional lymph node(s) be documented, as well as the choice of the Pediatric Regional Nodes code in a related STAGE text field on the abstract. Information on regional node status can be found on physical exam, scans, and pathology reports.

9625: Pediatric Mets

Item Length: 2
NAACCR Item #: 9625
XML Parent-NAACCR ID: pediatricMets
Active years: 2024+

Description

Pediatric Mets is part of the Pediatric Staging data collection system and is used to classify the distant site(s) of metastatic involvement at time of diagnosis**.** See also Pediatric Primary Tumor **[NAACCR Data item #9623]** and Pediatric Regional Nodes **[NAACCR Data item #9624]**. Effective for cases diagnosed 1/1/2024 and forward.

Rationale

Pediatric Mets is used to calculate Derived Pediatric M (when applicable) **[NAACCR Data item #9604].** Derivation will occur at the level of the central registry.

See the most current version of SEER*RSA (Pediatric Staging tab) for rules and site-specific codes and coding structures.

Code	Description
00	No distant metastasis
	SCHEMA-SPECIFIC CODES WHERE NEEDED
70	Distant metastasis, NOS
88	Not applicable
99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only

Coding Instructions

1. Code Pediatric Mets 00 (negative) instead of 99 (unknown) when **ALL** three of the following conditions are met:
   a. There is no mention of mets in the physical examination, pre-treatment diagnostic testing, or surgical exploration

   b. The patient has localized disease

   c. The patient receives what would be the standard treatment to the primary site (treatment appropriate to the stage of disease as determined by the physician), or patient is offered usual treatment but refuses it

   These guidelines apply only to localized cancers. Assign code 99 when there is reasonable doubt that the tumor is localized and there is no mention of absence or presence of metastatic disease.

   - ***Note:*** *This instruction is different than what is given for AJCC, EOD, and Summary Stage, which all have instructions that a tumor is determined to have no metastatic disease unless proven otherwise. These have not changed*
     - *This means it will be possible to code a case as "no metastatic involvement" in AJCC, EOD, and Summary Stage, and code 99 for unknown in Pediatric Mets*

5. **Noncontiguous/Discontinuous or hematogenous metastases:** Distant metastasis known at the time of diagnosis is coded in Pediatric Mets. In other words, when the patient was diagnosed, tumor had already spread indirectly (through vascular or lymph channels) to distant lymph nodes or to site(s) distant from the primary site. Refer to the individual schemas for detailed instructions.

6. **Positive pathological findings take priority over clinical findings.**

    a. Assign the highest applicable code for metastasis at diagnosis pathologically (based on pathology report), when available
        i. Not every metastatic site may be biopsied; however, for purposes of coding this data item, each metastatic site, whether confirmed clinically or pathologically, should be included, which may mean that clinical evidence would take priority over pathological

    b. If there is no applicable pathology or the pathology does not show metastasis, code Pediatric Mets based on clinical findings. Imaging takes precedence over physical examination.

7. Not all possible metastatic sites are listed in each of the schemas. If there is confirmed metastasis of a site that is not listed, assign the highest code as described below.

    b. For schemas that have only codes 10 (distant lymph nodes) and 70 (all other mets), code 70 is to be used for all mets (except distant lymph nodes only)

    c. For schemas where there are additional codes, use the highest code before code 70 when mets are present that are not specified in any of the other codes. Code 70 in these cases should only be used when the only information is "distant metastasis, NOS," and there is no documentation regarding the specific metastases

        i. For schemas where there are multiple distant site codes and the specific mets is not described, use the code that includes "other specific metastasis."
        ii. There will be enough information to code the numerically lower, but more specific, Pediatric Mets code when the location of the metastases is documented in the chart or abstract.

8. **Neoadjuvant (preoperative) therapy:** If the patient receives neoadjuvant (preoperative) systemic therapy (chemotherapy, immunotherapy) or radiation therapy, code the clinical information description that identifies the most extensive metastasis. If the post-neoadjuvant surgery shows additional or more extensive metastasis, code Pediatric Mets based on the post-neoadjuvant information. If the clinical and pathological information are the same, code mets based on the clinical information.

9. **Code 99**
    a. Assign code 99 when there is no information on metastatic disease and the primary tumor is not localized OR

    b. Death Certificate only (DCO) case

10. **Document choice of Pediatric Mets code in text.** It is strongly recommended that the positive and negative assessment of distant lymph nodes and/or distant metastasis be documented, as well as the choice of the Pediatric Mets code in a related STAGE text field on the abstract. Information on distant mets can be found mostly in Physical Exam and Scans.

9607: Derived Pediatric T

Item Length: 3
NAACCR Item #: 9607
XML Parent-NAACCR ID: derivedPediatricT
Active years: 2024+
New Data Item for Diagnosis Year 2024 and forward. Derived in Central registry only.

Description

This item stores the derived Pediatric T value from Pediatric Primary Tumor **[NAACCR Data item #9623]** and other fields (when applicable). Effective for cases diagnosed 1/1/2024 and forward.

Rationale

Derived Pediatric T can be used to evaluate disease spread at diagnosis, treatment patterns and outcomes over time.

9605: Derived Pediatric N

Item Length: 3
NAACCR Item #: 9605
XML Parent-NAACCR ID: derivedPediatricN
Active years: 2024+

New Data Item for Diagnosis Year 2024 and forward. Derived in Central registry only.

Description

This item stores the derived Pediatric N value from Pediatric Regional Nodes **[NAACCR Data item #9624]** and other fields (when applicable). Effective for cases diagnosed 1/1/2024 and forward.

Rationale

Derived Pediatric N can be used to evaluate disease spread at diagnosis, treatment patterns and outcomes over time.

9604: Derived Pediatric M

Item Length: 3
NAACCR Item #: 9604
XML Parent-NAACCR ID: derivedPediatricM
Active years: 2024+

New Data Item for Diagnosis Year 2024 and forward. Derived in Central registry only.

Description

This item stores the derived Pediatric M value from Pediatric Mets **[NAACCR Data item #9625]** and other fields (when applicable). Effective for cases diagnosed 1/1/2024 and forward.

Rationale

Derived Pediatric M can be used to evaluate disease spread at diagnosis, treatment patterns and outcomes over time.

9606: Derived Pediatric Stage Group

Item Length: 3
NAACCR Item #: 9606
XML NAACCR ID: derivedPediatricStageGroup
Active years: 2024+

Derived Pediatric Stage Group is derived using the Pediatric Stage Data Items (Derived Pediatric T [**NAACCR Data item #9607]**, Derived Pediatric N **[NAACCR Data item #9605]** and Derived Pediatric M **[NAACCR Data item #9604]**) algorithm. Other data items may be included in the derivation process. Effective for cases diagnosed 1/1/2024 and forward.

Rationale

Derived Pediatric Stage Group can be used to evaluate disease spread at diagnosis, treatment patterns and outcomes over time.

9628: Toronto T

Item Length: 3
Provisional NAACCR Item #: 9628
Provisional XML Parent-NAACCR ID: torontoT
Active years: 2025+
New Data Item for Diagnosis Year 2025 and forward. Derived in Central registry only.

Description

This item stores the Toronto T value derived from coded fields the Toronto Stage algorithm (when applicable). Effective for cases diagnosed 1/1/2025 and forward.

Rationale

Toronto T can be used to evaluate disease spread at diagnosis, treatment patterns and outcomes over time.

9629: Toronto N

Item Length: 3
Provisional NAACCR Item #: 9629
Provisional XML Parent-NAACCR ID: torontoN
Active years: 2025+

New Data Item for Diagnosis Year 2025 and forward. Derived in Central registry only.

Description

This item stores the Toronto N value derived from coded fields the Toronto Stage algorithm (when applicable). Effective for cases diagnosed 1/1/2025 and forward.

Rationale

Toronto N can be used to evaluate disease spread at diagnosis, treatment patterns and outcomes over time.

9630: Toronto M

Item Length: 3
Provisional NAACCR Item #: 9630
Provisional XML Parent-NAACCR ID: torontoM
Active years: 2025+

New Data Item for Diagnosis Year 2025 and forward. Derived in Central registry only.

Description

This item stores the Toronto M value derived from coded fields the Toronto Stage algorithm (when applicable). Effective for cases diagnosed 1/1/2025 and forward.

Rationale

Toronto M can be used to evaluate disease spread at diagnosis, treatment patterns and outcomes over time.

9631: Toronto Stage Group

Item Length: 3
Provisional NAACCR Item #: 9631
Provisional XML Parent-NAACCR ID: torontoStageGroup
Active years: 2025+

Toronto Stage Group is derived using the Toronto Stage Data Items (Toronto T [**NAACCR Data item #9628]**, Toronto N **[NAACCR Data item #9629]** and Toronto M **[NAACCR Data item #9630]**) algorithm. Other data items may be included in the derivation process. Effective for cases diagnosed 1/1/2025 and forward.

Rationale

Toronto Stage Group can be used to evaluate disease spread at diagnosis, treatment patterns and outcomes over time.

Prognostic Factors General Instructions

Timing for collection of SSDIs

The SSDIs are to be collected during the initial diagnosis, work up, and first course of treatment.

Consult Reports

If a report is sent out for consultation and the results are different than the original report, record the results from the consultation.

General Definitions and Format of SSDI Codes

Not applicable: This code is to be used ONLY when the data item is relevant for the case and the standard setter does not require the data item. Not applicable codes ALWAYS end in an 8 but will differ depending on the length of the data item.

Note: "Not applicable" is not available for schema discriminators or data items which are required for staging.

***Examples*:**

- n-MYC amplification. This is a 1-digit field. "Not applicable" is **8**
- White Blood Cell Count. This is a 7-digit field including the decimal point. "Not applicable" is **XXXXX.8**

It is important to review each data item carefully to determine how the "not applicable" code is formatted.

Unknown: Unknown codes ALWAYS end in a 9 but will differ depending on the length of the data item. The unknown code includes

- Test/evaluation/assessment **not** done or UNKNOWN if done

Examples:

- n-MYC amplification. This is a 1-digit field. "Unknown" is **9**
- White Blood Cell Count. This is a 7-digit field including the decimal point. "Unknown" is **XXXXX.9**

"Cannot be determined" on the pathology report. For some data items, this is a selection box on the College of American Pathologists (CAP) protocol. "Cannot be determined" is primarily used when a tissue specimen is not adequate for testing.

- If the pathology report includes "cannot be determined," code unknown

"Not identified." For some data items, this is a selection box on the CAP protocol. This means that the pathologist has looked for it and it is not present. This is not the same thing as looking for it in the medical record and not finding it (this would be "not documented in the medical record")

- For some SSDIs, "not identified," may be a specific code description
- If the pathologist stated, "not identified," and the SSDI does not include a specific code for not identified, code to negative

Death Certificate Only (DCOs) cases

For DCOs, the applicable SSDIs (except for applicable Schema Discriminators) may be blank.

- *Note*: This instruction is for central registries only.

Source Documents

Source documents are suggested for some data items as the most likely sources of information.

- If no source document is suggested, use any information provided in the medical record
- If a pathology report is suggested that document includes
    - Addenda or revisions to the report
    - Gross or microscopic description
    - Synoptic reports
    - CAP protocol provided by the pathologist

It is important to review each data item carefully to determine where the information can be found. For some data items, the information is based on imaging or some other type of clinical exam. Other data items are based on pathological findings from a surgical resection or molecular reports based on the tissue (usually added as an addendum to the pathology report).

Rounding Rules

SSDIs follow the standard definitions for rounding. These general rules can be followed for most SSDIs where lab values or percentages are recorded. All SSDIs that have lab values, percentages or measurements are set up to record in the 10ths (one digit after the decimal point). If a lab value, percentage, or measurement is recorded in 100ths (two digits after the decimal point), then the last digit must be rounded.

The general rounding rules are:

- If digit is 0-4, round down
- If digit is 5-9, round up

Example

- White Blood Cell Count is 85.25
    - Since the last digit is 5, round up and record 85.3
- White Blood Cell Count is 72.24
    - Since the last digit is 4, round down and record 72.2

Recording values when "less than" or "greater than" are used

Record the value as **one less** than stated when a value is reported as "less than X," and as **one more** than stated when a value is reported as "more than X." **One less** or **one more** may refer to a whole number (1), or a decimal (0.1), depending on the code structure of the field.

Example 1: White Blood Cell Count < (less than) 5. Record 4.9.

Example 2: White Blood Cell Counts > (greater than) 20. Record 20.1.

General Rules for Recording Laboratory Values

Laboratory values refer to any tests that are based on blood, urine, ascites, or spinal fluid. Most of these are based on blood.

Do not apply these rules to SSDIs that are based on tissue; see General Rules for Recording Tests Based on Solid Tissue.

Follow the below guidelines for recording laboratory values:

- All laboratory values must be done no earlier than approximately three months before diagnosis
- Only record test results obtained before any cancer-directed treatment is given (neoadjuvant therapy or surgical), unless instructions for a specific laboratory test state otherwise
- Record the highest laboratory value if multiple laboratory tests results are available, unless instructions for a specific laboratory test state otherwise

General Rules for Entering Laboratory Values and Other Measurements

Lab values and other measurements that are not integers (whole numbers) and are reported as continuous variables (not categories or ranges) will be recorded to a single decimal place with an explicit decimal point.

There must always be a numeral or the letter 'X' immediately before the decimal point and a numeral after the decimal point, which will be in the next-to-last character position in the field. The entered value must be right-justified in the field and padded with spaces to the left if necessary to fill the field.

Users' software will usually justify and pad the value automatically for the registrar.

In addition to the actual values, codes are defined for situations such as value unknown; test done but results not in chart; and other special cases. Sometimes codes will be provided for when a value is expressed as "at least" some value.

- These may be needed, for *example*, in the measurement of tumor size or thickness when the tumor has been transected and the actual size cannot be determined. These codes will begin with one or more 'X's.

When a value in the medical record does not provide the expected decimal digit, i.e., it is expressed as a whole number, then enter the value followed by a decimal point and a zero.

Examples for a 6-Character Lab Value

Value in Record	Data Item Coded as
0.0	0.0
0	0.0
.1	0.1
11.0	11.0
11.1	11.1
11	11.0
111.1	111.1
1111.1	1111.1

General Rules for Recording Tests Based on Solid Tissue

Priority Order for SSDIs

- Addendums or amendments (corrections that are not incorporated into the initial synoptic report, results from molecular studies, and including CAP Cancer Protocol)
- Synoptic report (including CAP Cancer Protocol)
- Pathology report: final diagnosis
- Physician statement

For these SSDIs, a microscopic evaluation (tissue examination) is required.

- If no microscopic evaluation (biopsy, surgical resection), code the SSDI to the unknown value

General Rules versus SSDI specific rules

- Unless instructions for a specific tissue test state otherwise, record the highest value (positive versus negative, or actual numerical value) obtained from any tissue-based examination (biopsy, surgical resection, bone marrow biopsy).

Pediatric Stage Data Items and SSDIs by Pediatric ID

The next section contains the schema specific information for Pediatric Primary Tumor, Pediatric Regional Nodes, Pediatric Mets, and the applicable SSDIs.

1a1: Acute Lymphoblastic Leukemia

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C000-C809	9811-9819, 9837	0-39	3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Acute Lymphoblastic Leukemia, Toronto Staging is based on the presence or absence of CNS Involvement, which is collected in Pediatric Mets.

9623: Pediatric Primary Tumor

- Not applicable (code 888)

9624: Pediatric Regional Nodes

- Not applicable (code 888)

1a1: Acute Lymphoblastic Leukemia

9625: Pediatric Mets

Coding Instructions and Codes

Note 1: This field records the clinically or cytologically confirmed involvement of the Central Nervous System at diagnosis or during the initial workup of Acute Lymphoblastic Leukemia (ALL) in pediatric patients. CNS involvement at diagnosis or at relapse is a significant cause of treatment failure and treatment-related morbidity.

- Use the following resources to find information on CNS involvement: imaging, laboratory reports, CSF lactate dehydrogenase (LD, LDH) test

Note 2: Physician statement of CNS Involvement can be used to code this data item.

Note 3: Criteria for CNS Involvement

- Clinical signs of CNS involvement
  - Radiologic evidence of intracranial, intradural mass
  - Cranial nerve palsy (e.g., facial weakness, ptosis), brain/eye involvement or hypothalamic syndrome
  - *Note*: Extra-ocular orbital masses, severe headaches, and eye swelling (in the absence of signs of cranial nerve involvement) are **not** sufficient to constitute clinical evidence of CNS involvement
- The presence of blasts in the cerebrospinal fluid (CSF)
  - Cytospin is used to determine the presence or absence of blasts in the CSF.
    - If Cytospin is not documented, then the presence of blasts cannot be determined. CNS involvement would be unknown (code 9) unless there are clinical statements of CNS Involvement **OR the physician documents a CNS stage**
  - If Cytospin is documented and there is no mention of blasts, code blasts as absent
  - If blasts are referred to as "occasional" or "seen" or similar wording, code blasts as present
- Red and white blood cell counts from the cerebrospinal fluid (RBC CSF and WBC CSF)
  - Lumbar punctures are the most common way to collect this information
  - If RBC <1/ul, record as RBC = 0
  - If WBC <1/uL, record as WBC = 0

Code	Description	SS2018
10	CNS1 • No clinical signs of CNS involvement AND • No blasts in CSF	D

20	CNS2
	• No clinical signs of CNS involvement AND blasts in CSF AND
	  ○ WBC < 5/ul CSF
	OR
	• No clinical signs of CNS involvement OR blasts in CSF AND
	  ○ WBC >= 5/ul CSF and
	  ○ RBC >= 10/ul CSF AND
	  ○ WBC/RBC in CSF <= 2x WBC/RBC in blood
		D

Code	Description	SS2018

30	CNS3
	• Clinical signs of CNS involvement
	OR
	• Blasts in CSF and WBC >= 5/ul CSF AND EITHER
	  ○ RBC < 10/ul CSF OR
	  ○ RBC >= 10/ul CSF and WBC/RBC in CSF > 2x WBC/RBC in blood
		D

99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only	D

1a1-Acute Lymphoblastic Leukemia

9621: White Blood Cell Count

Item Length: 7
NAACCR Item #: 9621
XML NAACCR ID: whiteBloodCellCount
Active years: 2024+
Pediatric Schema(s):

- 1a1: Acute Lymphoblastic Leukemia (ALL)

Description

White Blood Cell Count (WBC) will record the actual lab value prior to treatment.

Rationale

This is part of the National Childhood Cancer Registry (NCCR) project to collect more specific information on pediatric patients. Registries part of the NCCR will start collection on specific pediatric data items with 2024 diagnoses.

Coding Instructions and Codes

Source documents: laboratory reports (white blood cell count)

Note 1: This SSDI is effective for diagnosis years 2024+

- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: Physician statement of WBC (White Blood Cell Count) Pretreatment Lab Value can be used to code this data item when no other information is available.

- Record the lab value of the highest WBC test result documented in the medical record **prior to treatment**. The lab value may be recorded in a lab report, history and physical, or clinical statement in the pathology report
- Record to the nearest cells per microliter (cells/uL) the highest WBC lab value documented in the medical record **prior to treatment**
- A known lab value takes priority over codes XXXXX.2-XXXXX.4
    - The lab value takes priority even if the physician documents the interpretation
      Example: Patient noted to have a WBC count of 12,000 cells/mm^3. Physician notes that the value is elevated
      Code 12,000 cells/mm^3 instead of XXXXX.4 (elevated)

Note 3: The measurement used for this data item is "cells/mm3", which is the same as "cells/uL". If your facility has a different measurement, here are examples of the conversions to cells/m3.

- *Examples:*
    - Reported as 3650 cells/uL. Record as 3650.0
    - Reported as 3.55 k/uL. Record as 3550.0 (multiple x 1000)
    - Reported as 4.25 10^3/uL. Record as 4250.0 (multiple x 1000)
    - Reported as 6.35 K/mm^3. Record as 6350.0 (multiple x 1000)

Code	Description
0.0-99999.9	0.0-99999.9 microliter (cells/mm^3) (Exact value to nearest tenth in (cells/mm^3)
XXXXX.1	100,000 or greater (cells/mm^3)

Code	Description
XXXXX.2	Lab value not available, physician states WBC is low
XXXXX.3	Lab value not available, physician states WBC is normal
XXXXX.4	Lab value not available, physician states WBC is elevated/high
XXXXX.7	Test ordered, results not in chart
XXXXX.8	Not applicable: Information not collected for this case (If this information is required by your standard setter, use of code XXXXX.8 may result in an edit error)
XXXXX.9	Not documented in medical record WBC (White Blood Cell Count) Pretreatment Lab Value not assessed or unknown if assessed
<BLANK>	N/A - Diagnosis year is prior to 2024

2a: Hodgkin Lymphoma

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C000-C809	9650-9653, 9655, 9659, 9663	0-39	3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Hodgkin Lymphoma, Toronto Staging is based on the *Ann Arbor Staging Classification* and is collected in Pediatric Primary Tumor

- Internationally, the Ann Arbor Stage is still used for Lymphomas. Ann Arbor Stage is very similar to the Lugano Stage that has been collected in the US since 2018. The major difference between the two is that Lugano Stage has bulky disease, and Ann Arbor does not (**see Note 5** in Pediatric Primary Tumor)

2a: Hodgkin Lymphoma

9623: Pediatric Primary Tumor

Coding Instructions and Codes

Note 1: Lymphatic sites (nodal regions) are

- Lymph nodes (C770-C779)
- Waldeyer' s ring (tonsils) (C024, C090-C099, C111, C142)
- Spleen (C422)
- Thymus (C379)

Note 2: Use the AJCC definitions for lymph node regions (Chapter 79, Figure 79.1) to determine when single (code 100) or multiple (300-600) lymph node regions are involved. See also the Hematopoietic Manual, Appendix C, for definition of lymph node regions.

Note 3: Extralymphatic sites (extranodal regions) include all other sites (e.g., stomach, colon, lung, breast, nasopharynx).

Note 4: Any mention of the terms including fixed, matted, mass in the hilum, mediastinum, retroperitoneum, and/or mesentery, palpable, enlarged, shotty, lymphadenopathy are all regarded as involvement for lymphomas when determining appropriate code.

Note 5: For Hodgkin lymphoma, "Bulky disease" is defined as the ratio between the maximum diameter of the mediastinal mass and maximal intrathoracic diameter based on CT imaging in the Lugano classification. Bulk of other disease is defined as a mass greater than 10 cm. This is the only difference between Lugano Staging and Ann Arbor, which does not include "bulky disease."

- If there is mention of bulky disease without further involvement, code 300 or 400 for a nodal lymphoma and 400 for an extranodal lymphoma

Note 6: Clinical enlargement of the liver is not enough to indicate involvement. Involvement is indicated by diffuse uptake or mass lesion or abnormal liver function tests. Liver biopsy may be used to confirm equivocal involvement.

- Any involvement of liver (including primary liver lymphoma) is coded as 800

Note 7: Splenic involvement is based on splenomegaly and FDG-PET or CT scans that state diffuse update, solitary mass, miliary lesions, or enlargement of greater than 13 cm.

- A physician's statement of splenomegaly may be used
- FDG uptake in the spleen that is not diffuse is not enough to code as splenic involvement

Note 8: Lung involvement is indicated by pulmonary nodules or parenchymal involvement on FDG-PET or CT in the absence of other likely causes. Lung biopsy may be used to confirm equivocal involvement.

- Multifocal lung involvement is coded as 700 or 800 based on lung mets, code also "Mets at Dx-Lung" as 1

Note 9: Bone involvement (excluding bone marrow involvement, **see Note 11**) is indicated by avid lesions on FDG-PET. Bone biopsy may be used to confirm equivocal involvement.

- Bone involvement (except for bone primary lymphomas) is coded as 800. Code also "Mets at Dx-Bone" as 1. (**see Note 11** on how to code bone marrow involvement)

Note 10: Central nervous system (CNS) involvement is often suspected due to symptoms and can be confirmed by plain radiology, CT scan, or MRI. Cerebrospinal fluid (CSF) examination by flow cytometry may be done. CNS involvement may be the result of soft tissue disease representing extension from bone metastasis or parenchymal brain disease.

- CNS involvement (except for CNS primary lymphomas) is coded as 800. Code also "Mets at Dx-Brain" as 1

- CSF involvement is coded as 800. Code also "Mets at Dx-Other" as 1

Note 11: Peripheral blood involvement is assessed by an aspiration or peripheral blood smear.

- Primary site is coded to bone marrow (C421): Do not code "Met at Dx-Other" as 1
    - In cases where peripheral blood smear is not performed, but a physician's clinical assessment indicates peripheral blood involvement, the physician's clinical assessment can be used
    - If **ONLY** the peripheral blood is involved, code 750
    - If there is peripheral blood involvement **WITH** other involvement
        - Do not code primary site to C421, code to lymph nodes or organs involved
        - Pediatric Primary Tumor will based on the involvement of the lymph nodes or organs

Note 12: Bone marrow involvement is assessed by an aspiration and bone marrow biopsy.

- Bone marrow involvement is coded as 800. Do not code to "Mets at Dx-Bone" as 1
    - If only involvement is bone marrow, code primary site to bone marrow (C421), Pediatric Primary Tumor 800. Do not code "Mets at Dx-Other" as 1
    - If there is involvement of lymph nodes or organs AND bone marrow, code “Mets at Dx-Other” as 1
    - In cases where bone marrow biopsy/aspiration is not performed, but a physician's clinical assessment indicates bone marrow involvement, the physician's clinical assessment can be used

Note 13: See the data item *B symptoms* [NAACCR Data Item Number: #3812] to code the presence or absence of B symptoms.

Code	Description	SS2018
100	Nodal lymphomas • Single lymph node region involved • Involvement of multiple nodal chains in the SAME lymph node region	L

200	Extranodal lymphomas
	• Single extralymphatic site
	  ○ WITHOUT nodal involvement
	• Multifocal involvement (except multifocal lung involvement or any liver involvement, see code 800) of one extralymphatic organ/site
	  ○ WITHOUT nodal involvement (see code 400 for WITH nodal involvement)
		L

300	Nodal lymphomas • Two or more lymph node regions involved ○ SAME side of diaphragm WITH or WITHOUT bulky disease	RE

400	Nodal lymphomas
	• Contiguous extralymphatic extension from nodal/lymphatic site
	• WITH or WITHOUT involvement of other nodal regions
	  ○ on SAME side of diaphragm
	Extranodal lymphomas
	• Localized involvement of a single extralymphatic organ/site
	  ○ WITH involvement of its regional lymph node(s) OR
	  ○ WITH involvement of other lymph node(s) on the SAME side of the diaphragm
	
	WITH or WITHOUT bulky disease
		RE

Code	Description	SS2018
575	Nodal and Extranodal lymphomas • Involvement of lymph node regions on BOTH sides of the diaphragm ○ WITHOUT or UNKNOWN spleen involvement	D
600	Nodal and Extranodal lymphomas • Involvement of lymph node regions on BOTH sides of the diaphragm WITH spleen involvement ○ Includes involvement of lymph nodes ABOVE the diaphragm WITH spleen involvement	D
700	Diffuse or disseminated involvement (except multifocal lung involvement or any liver involvement, see code 800) of ONE OR MORE extralymphatic organ(s)/site(s) • WITH or WITHOUT nodal involvement Involvement of isolated extralymphatic organ in absence of involvement of adjacent lymph nodes, but in conjunction with disease in distant sites Multifocal involvement (except multifocal lung involvement or any liver involvement, see code 800) of one extralymphatic organ/site • WITH nodal involvement Noncontiguous extralymphatic organ involvement in conjunction with nodal disease (two or more sites involved)	D
750	Peripheral blood involvement ONLY	D
800	Diffuse or disseminated • Bone • Central nervous system (CNS) Any involvement of • Bone marrow • Cerebrospinal fluid (CSF) • Liver • Lung, multiple lesions (other than by direct extension in code 400) • Peripheral blood involvement WITH other involvement Distant metastasis, NOS	D
999	Unknown; extension not stated Tumor cannot be assessed Not documented in medical record Death Certificate Only	U

9624: Pediatric Regional Nodes

- Not applicable (code 888)

9625: Pediatric Mets

- Not applicable (code 88)

2a: Hodgkin Lymphoma

3812: B Symptoms

Item Length: 1
NAACCR Item #: 3812
XML NAACCR ID: bSymptoms
NAACCR Alternate Name: None
Active years: 2018+
Schema(s):

- 2a: Hodgkin Lymphoma (2024+)

Description

B symptoms refer to systemic symptoms of fever, night sweats, and weight loss which can be associated with both Hodgkin lymphoma (HL) and some non-Hodgkin lymphomas (NHL). The presence of B symptoms is a prognostic factor for some lymphomas.

Rationale

B symptoms is a Registry Data Collection Variable in AJCC. This data item was previously collected for Lymphomas, SSF #2. Registries part of the NCCR will start collection on specific pediatric data items with 2024+ diagnoses.

The stages of Hodgkin Lymphoma are classified as either A or B according to the absence or presence of defined constitutional symptoms. The stage group suffix for a patient without these systemic symptoms is "A," meaning absence of symptoms or asymptomatic; *for example*, Stage IIA. The stage group suffix for a patient with any of the symptoms listed below is "B," such as Stage IIIB. The symptoms are carefully defined.

- Fevers: Unexplained fever with temperature above 38 degrees centigrade or 101.5 degrees Fahrenheit.
- Night sweats: Drenching sweats (e.g., those that require change of bedclothes)
- Weight loss: Unexplained weight loss of more than 10% of the usual body weight in the 6 months prior to diagnosis.

Other symptoms, such as chills, pruritic, alcohol-induced pain, and fatigue, are not included in the A or B designation but are recorded in the medical record, as the reappearance of these symptoms may be a harbinger of recurrence. The designation A or B is not included in the revised staging of NHL in AJCC8, although clinicians are encouraged to record the presence of these symptoms in the medical record. The presence or absence of B symptoms may be collected in registries for both HL and NHL.

Coding Guidelines

- Code 0 when there is no evidence of B symptoms present, per physician or physical exam
- Code 1 when the physician states the patient has B symptoms
- Code 9 when
    - Not documented in the medical record
    - B symptoms not evaluated (assessed)
    - Unknown if B symptoms evaluated (assessed)

Additional Information

- **Source documents:** patient history, progress notes, consultant notes, other statements in medical record
- **Other names:** B symptoms; Fever: Palestine fever, hyperpyrexia, febrile response; sleep hyperhidrosis, nocturnal hyperhydrosis

- **Note**: This was previously required for staging under the Ann Arbor Staging Classification for Lymphomas. The Lugano Staging System does not require this for staging.
    - **Per AJCC 8th edition:** "The designation A or B is not included in the revised staging of NHL, although clinicians are encouraged to record the presence of these symptoms in the medical record."
    - If your physicians no longer record the B symptoms because of this change, code 9
- ***Change from Collaborative Stage v2 (CSv2):*** *In **CSv2**, if there was no mention of B symptoms, the registrar could assume that they were not present and code appropriately. For the SSDI, this assumption cannot be made. There must be a statement that B symptoms are not present to assign code 0.*

Coding Instructions and Codes

Note 1: Physician statement of B symptoms can be used to code this data item when no other information is available.

Note 2: Each stage should be classified as either A or B according to the absence or presence of defined constitutional symptoms, such as

- Fevers: Unexplained fever with temperature above 38 degrees C (100.4 F)
- Night sweats: Drenching sweats that require change of bedclothes
- Weight loss: Unexplained weight loss of more than 10% of the usual body weight in the six months prior to diagnosis

Note 3: Pruritus alone does not qualify for B classification, nor does alcohol intolerance, fatigue, or a short, febrile illness associated with suspected infections.

Note 4: Code 9 if there is no mention of B symptoms.

Code	Description
0	No B symptoms (asymptomatic) Classified as "A" by physician when asymptomatic
1	Any B symptom(s) Night sweats (drenching) Unexplained fever (above 38 degrees C) Unexplained weight loss (generally greater than 10% of body weight in the six months before admission) B symptoms, NOS Classified as "B" by physician when symptomatic
8	Not applicable: Information not collected for this case (If this item is required by your standard setter, use of code 8 will result in an edit error.)
9	Not documented in medical record B symptoms not assessed or unknown if assessed

2b2-2b4, 2c: Non-Hodgkin Lymphoma

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C000-C809	9591, 9671, 9673, 9678-9680, 9687-9691, 9695, 9698, 9699-9702, 9705, 9714-9719, 9724, 9735, 9737-9738, 9761-9762, 9765-9768, 9769, 9823, 9827, 9970-9971	0-39	3
C000-C809	9731-9732, 9734	0-39	3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For non-Hodgkin Lymphoma, Toronto Staging is based on the *St. Jude/Murphy Staging System*, which is a pediatric staging system for non-Hodgkin lymphoma (NHL) and is collected in Pediatric Primary Tumor.

- The definitions for StJude/Murphy Staging are different than the Lugano Staging or Ann Arbor, which has historically been used in the US to record Lymphoma stage

2b2-2b4, 2c: Non-Hodgkin Lymphoma

9623: Pediatric Primary Tumor

Coding Instructions and Codes

Note 1: Any mention of the terms including fixed, matted, mass in the hilum, mediastinum, retroperitoneum, and/or mesentery, palpable, enlarged, shotty, lymphadenopathy are all regarded as involvement for lymphomas when determining appropriate code.

Note 2: St. Jude/Murphy staging is based primarily on the primary site where certain primary sites or sites of involvement may only be assigned specific codes. The list below provides guidance on which codes to use based on the primary site and/or other involvement.

- Codes based on primary site
  - Abdomen (C239, C240-C249, C250-C259, C422, C493-C494, C480-C488, C649, C659, C669, C762, C772), then only code 400 may be assigned
  - GI Tract (C160-C218, C260-C269), then only codes 200 or 400 may be assigned
  - Paraspinal or epidural sites (C470-C479), then only code 400 may be assigned
  - Primary intrathoracic tumors (mediastinal, hilar, pulmonary, pleural, or thymic) (C340-C349, C379, C381-C388, C771, then only code 400 may be assigned
- Codes based on involvement
  - Code 600 for liver involvement (including primary site C220)
  - Code 800 for bone marrow (including primary site C421), or peripheral blood involvement
  - Code 800 for CNS involvement (including primary sites C700-C729)
- ALL sites except those noted above, codes 100, 300, 500-800, 999 may be assigned based on involvement

Note 3: Extensive disease (code 400) typically exhibits spread to para-aortic and retroperitoneal areas by implants and plaques in mesentery or peritoneum, or by direct infiltration of structures adjacent to the primary tumor. Ascites may be present, and complete resection of all gross tumour is not possible.

Note 4: Code 888 for histologies 9731, 9732, and 9734

- St Jude Staging is not applicable for Plasmacytomas (9731, 9734) and Plasma Cell Myeloma (9732)

Code	Description	Ped Stage
100	Involvement of a single extranodal/extralymphatic tumor mass or nodal/lymphatic area *Excludes the following primary sites:* • Abdomen (See code 400) • Bone marrow (See code 800) • CNS (See code 800) • GI Tract (See code 200) • Liver (See code 600) • Paraspinal or epidural sites (See code 400) • Primary intrathoracic tumors (See code 400)	I
200	A completely resected primary gastrointestinal tract tumor (C160-C218, C260-C269) • WITH or WITHOUT involvement of associated **mesenteric nodes only** (for other lymph node involvement, see code 400)	II

Code	Description	Ped Stage

300	Any of the following (excluding primary sites in codes 200, 400, 600, 800)
	• A single extranodal/extralymphatic tumor with regional node involvement
	• Two or more nodal/lymphatic areas on the SAME side (either above or below) of the diaphragm or
	• Two or more single extranodal/extralymphatic tumors
	  ○ WITH or WITHOUT regional node involvement **AND**
	  ○ On the SAME side (either above or below) of the diaphragm
		II

400	Any of the following
	• Abdominal tumors
	  ○ Includes extensive (unresectable) primary intraabdominal disease
	• Gastrointestinal tumors
	  ○ WITH lymph node involvement other than mesenteric OR
	  ○ Unresectable gastrointestinal tract tumor
	• Paraspinal or epidural tumors regardless of other tumor sites
	• Primary intrathoracic tumors (mediastinal, hilar, pulmonary, pleural, or thymic)
		III

500	Extranodal/extralymphatic OR nodal/lymphatic tumors • On BOTH sides (above and below the diaphragm)	III
600	Liver involvement, including primary site (C220) • WITH or WITHOUT lymph node involvement	III
800	CNS involvement, including primary site (C700-C729) • Single, multifocal, or multiple CNS tumors, or CNS tumor WITH lymph node involvement • Cranial nerve palsy that cannot be explained by extradural lesions, OR • Blasts morphologically identified in CSF • In the absence of a CSF tumor mass and cranial nerve palsy, a CSF report is required to confirm or exclude CNS involvement AND/OR • Bone marrow involvement (including primary site bone marrow, C421) • Peripheral blood involvement WITH or WITHOUT other involvement	IV
888	Not applicable: histologies 9731, 9732, 9734	NA
999	Not documented in medical record St Jude Staging System not assessed Death certificate only	U

9624: Pediatric Regional Nodes

- Not applicable (code 888)

9625: Pediatric Mets

- Not applicable (code 88)

3a: Ependymoma

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C710-C729	9383, 9391-9394, 9396	All ages	0, 1, 3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Ependymoma, Toronto Staging is based on the presence or absence of mets based on the *Chang M* definition for metastases within the CNS at diagnosis and is collected in Pediatric Mets.

- Pediatric Primary Tumor is collected for surveillance purposes

3a: Ependymoma

9623: Pediatric Primary Tumor

Coding Instructions and Codes

Note 1: Pediatric Primary Tumor for Ependymomas is coded **only for single tumors confined to the primary site** (see code 150) or a **single** tumor crossing the midline without extension to adjacent structures (see code 250).

- Code 999 if there are multiple tumors in the brain
- The presence of multiple tumors is recorded in Pediatric Mets

Note 2: Benign (/0) or Borderline (/1) tumors are **always coded to 050** regardless of size or extension to adjacent sites

Note 3: A midline shift is not the same thing as crosses/crossing the midline

- Code 150 if you have a single tumor confined to the primary site with a midline shift that is not extending into adjacent structures (see Note 4).

Note 4: Direct or contiguous extension to an adjacent site is collected in Pediatric Mets.

- If the only information available is extension to an adjacent site, code Pediatric Primary Tumor 999 and assign the appropriate Pediatric Mets code
- The following adjacent structures/sites are collected in Pediatric Mets (see code 25 for all except circulating cells in CSF (code 15))
    - Adjacent connective/soft tissue
    - Adjacent muscle
    - Bone
    - Circulating cells in cerebral spinal fluid (CSF)
    - Major blood vessel(s)
    - Meninges (e.g., dura)
    - Multiple/multifocal tumors
    - Nerves (cranial, NOS)
    - Ventricular system

Code	Description	SS2018 T
050	Benign or borderline brain	B
150	All sites • Single tumor confined to the primary site with no invasion or seeding to other structures Confined to site of origin, NOS Localized, NOS	L
250	Single tumor confined to the primary site that crosses/crossing the midline • WITHOUT invasion of adjacent structures (see Note 4)	RE
800	No evidence of primary tumor	U
999	Unknown; extension not stated Multiple tumors (See Note 1) Single tumor with extension to an adjacent site (see Note 4) Primary tumor cannot be assessed Not documented in medical record Death Certificate Only	U

3a: Ependymoma

9624: Pediatric Regional Nodes

- Not applicable (code 888)

9625: Pediatric Mets

Coding Instructions and Codes

Note 1: Use code 70 when the only information is "distant metastasis, NOS," and there is no documentation regarding the specific metastases.

- If there are specific metastasis documented that are not listed in codes 15, 25, or 35, or 45, assign code 45 for "other specified distant metastasis."

Note 2: Code 00 for benign (behavior /0) and borderline (behavior /1) tumors.

Note 3: The following adjacent structures/sites, by direct or contiguous extension, are coded to 35.

- Adjacent connective/soft tissue
- Adjacent muscle
- Bone
- Circulating cells in cerebral spinal fluid (CSF)
- Major blood vessel(s)
- Meninges (e.g.; dura)
- Multiple/multifocal tumors
- Nerves (cranial, NOS)
- Ventricular system

Note 4: Leptomeningeal metastases, also known as carcinomatous meningitis and meningeal carcinomatosis, refers to the spread of malignant cells through the CSF space. These cells can originate from primary CNS tumors (e.g., in the form of drop metastases), as well as from distant tumors that have metastasized via hematogenous spread (code 35).

Code	Description	SS2018 M
00	No visible disease on imaging (MRI brain and spine) beyond primary site of disease AND no tumor cells into the cerebrospinal fluid (CSF)	None
15	Tumor cells in the CSF • Circulating cells in cerebral spinal fluid (CSF)	D

Code	Description	SS2018 M
25	Intracranial spread beyond a single lesion All sites • Bone (skull) (see code 45 for other bone involvement) • Major blood vessel(s) • Meninges (e.g., dura) • Multiple/multifocal tumors • Nerves (cranial, NOS) • Tumor invades or encroaches upon ventricular system Brain tumors (C700, C710-C719) • Anterior cranial fossa • Brain stem • Cerebellum • Cerebrum (cerebral hemisphere) • Contralateral hemisphere • Hypothalamus • Middle cranial fossa • Pallium • Posterior cranial fossa • Suprasellar brain • Tapetum • Thalamus CNS tumors (C701, C709, C720-C729) • Adjacent connective tissue • Adjacent muscle • Brain for cranial nerve tumor(s) • Sphenoid and frontal sinuses(skull) Pineal Gland (C753) • Adjacent connective/soft tissue • Cavernous sinus • Infratentorial and central brain	D
35	Visible metastasis in spine OR Visible metastasis in cervicomedullary (junction) Metastasis within CNS and CSF pathways • Carcinomatous meningitis • Drop metastasis • Leptomeningeal metastases • Meningeal carcinomatosis	D

Code	Description	SS2018 M
45	Extra-neural metastasis All Sites • Blood • Bone (other than skull) (see code 25 for skull) • Bone marrow • Carcinomatosis • Distant lymph nodes, NOS • Further contiguous extension • Other specified metastasis Brain tumors (C700, C710-C719) • Nasal cavity • Nasopharynx • Other direct extension outside CNS • Posterior pharynx CNS tumors (C701, C709, C720-729) • Eye	D
70	Distant metastasis, NOS	D
99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only	None

3b: Astrocytoma

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C700-C729	9380, 9384, 9400-9411, 9420-9424, 9440-9442, 9445	0-39	0, 1, 3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Astrocytoma, Toronto Staging is based on the presence or absence of mets and is collected in Pediatric Mets.

- Pediatric Primary Tumor is collected for surveillance purposes

3b: Astrocytoma

9623: Pediatric Primary Tumor

Coding Instructions and Codes

Note 1: The tentorium cerebelli is an extension of the dura mater that separates the cerebellum from the inferior portion of the occipital lobes. The location of the tumor above or below the tentorium can help in determining the type of tumor; also, most adult brain tumors are supratentorial, and most pediatric brain tumors are infratentorial. In the following list, note that ICD-O-3 codes C710 and C719 include both supratentorial and infratentorial subsites.

- The following subsites are **Infratentorial**
    - All subsites for codes C716-C717
    - Hypothalamus (C710)
    - Pallium (C710)
    - Posterior cranial fossa (C719)
    - Thalamus (C710)
- The following subsites are **Supratentorial**
    - All subsites for codes C711-C715
    - Primary site C710 (excluding hypothalamus, pallium, thalamus)
    - Anterior cranial fossa (C719)
    - Corpus callosum (C718)
    - Middle cranial fossa (C719)
    - Tapetum (C718)
    - Suprasellar (C719)

Note 2: Benign (/0) or Borderline (/1) tumors are **always coded to 050** regardless of size or extension to adjacent sites.

Note 3: A midline shift is not the same thing as crossing the midline (code 500).

- Documentation must state “crosses the midline”

Note 4: Discontiguous spread, or "drop metastasis" are coded in Pediatric Mets.

Code	Description	SS2018 T
050	Benign or borderline brain tumor	B

100	Brain (C700, C710-C719)
	
	Infratentorial tumor confined to
	• Brain stem or meninges of brain stem (one side)
	  ○ Medulla oblongata
	  ○ Midbrain (mesencephalon)
	  ○ Pons
	• Cerebellum or meninges of cerebellum (one side or midline)
	  ○ Lateral lobes
	  ○ Median lobe of cerebellum
	  ○ Vermis
	• Hypothalamus
	• Pallium
	• Thalamus
	
	Infratentorial tumor
	• Both cerebellum and brain stem involved with tumor on one side
	
	Supratentorial tumor confined to
	
	• Cerebral hemisphere (cerebrum) or meninges of cerebral hemisphere (one side)
	  ○ Frontal lobe
	  ○ Occipital lobe
	  ○ Parietal lobe
	  ○ Temporal lobe
	
	Confined to ventricles
	Tumor invades or encroaches upon ventricular system
	
	Confined to brain, NOS
	Confined to meninges, NOS
		L

200	CNS Other (C701, C709, C720-C729) Confined to tissue or site of origin Localized, NOS	L

Code	Description	SS2018 T
500	Brain (C700, C710-C719) Tumor crosses the midline Tumor invades • Bone (skull) (see code 700 for other bone involvement) • Contralateral hemisphere • Corpus callosum (including splenium) • Major blood vessel(s) • Meninges (e.g., dura) • Nerves (cranial, NOS) • Spinal cord/canal Supratentorial tumor extends infratentorially to involve • Brain stem • Cerebellum • Hypothalamus • Pallium • Posterior cranial fossa • Thalamus Infratentorial tumor extends supratentorially to involve • Anterior cranial fossa • Cerebrum (cerebral hemisphere) (excluding hypothalamus, pallium, thalamus) (see code 100 for hypothalamus, pallium, thalamus) • Middle cranial fossa • Suprasellar brain • Tapetum	RE
600	CNS Other (C701, C709, C720-C729) Adjacent connective/soft tissue Adjacent muscle Bone (skull) (see code 700 for other bone involvement) Brain for cranial nerve tumor(s) (see code 700 for sites other than cranial nerve tumors) Major blood vessel(s) Meningeal tumor infiltrates nerve Nerve tumor infiltrates meninges (dura) Sphenoid and frontal sinuses (skull)	RE

Code	Description	SS2018 T
700	Brain (C700, C710-C719) Circulating cells in cerebral spinal fluid (CSF) Bone other than Skull (see code 500 for skull) Nasal cavity Nasopharynx Other direct extension outside CNS Posterior pharynx Further contiguous extension	D
750	CNS Other (C701, C709, C720-C729) Bone other than skull (see code 400 for skull) Brain except for cranial nerve tumor(s) (see code 600 for cranial nerve tumors) Eye Further contiguous extension	D
800	No evidence of primary tumor	U
999	Unknown; extension not stated Primary tumor cannot be assessed Not documented in medical record Death Certificate Only	U

9624: Pediatric Regional Nodes

- Not applicable (code 888)

3b: Astrocytoma

9625: Pediatric Mets

Coding Instructions and Codes

Note 1: Code 00 for benign (behavior /0) and borderline (behavior /1) tumors.

Note 2: Leptomeningeal metastases, also known as carcinomatous meningitis and meningeal carcinomatosis, refers to the spread of malignant cells through the CSF space. These cells can originate from primary CNS tumors (e.g., in the form of drop metastases), as well as from distant tumors that have metastasized via hematogenous spread (code 70).

Code	Description	SS2018 M
00	No distant metastasis	None
10	Distant lymph node(s)	D
70	Metastasis within CNS and CSF pathways • Carcinomatous meningitis • Drop metastasis • Leptomeningeal metastases • Meningeal carcinomatosis Metastasis outside the CNS Extra-neural metastasis Carcinomatosis Distant metastasis WITH or WITHOUT distant lymph node(s) Distant metastasis, NOS	D
99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only	None

3b: Astrocytoma

3940: BRAF Mutational Analysis

Item Length: 1
NAACCR Item #: 3940
XML NAACCR ID: brafMutationalAnalysis
Active years: 2018+
Schema(s):

Description

The BRAF oncoprotein is involved in transmitting cell growth and proliferation signals from KRAS and NRAS. The BRAF V600E mutation is associated with poorer prognosis and predicts lack of response to anti-EGFR therapies.

Rationale

This is part of the National Childhood Cancer Registry (NCCR) project to collect more specific information on pediatric patients. Registries part of the NCCR will start collection on specific pediatric data items with 2024+ diagnoses.

Coding Instructions and Codes

Possible sources of information: molecular pathology reports (may be addendum to original pathology report)

Note 1: This SSDI is effective for diagnosis years 2024+
- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: Physician statement of BRAF can be used to code this data item when no other information is available.

Note 3: BRAF is a gene which belongs to a class of genes known as oncogenes. When mutated, oncogenes have the potential to cause normal cells to become cancerous. Studies suggest that BRAF gene mutations are often present in Astrocytomas. The most common BRAF mutations are
- BRAF V600E (c.1799T>A) mutation
- *KIAA1549*:*BRAF* gene fusion

Note 4: The most common testing methods for BRAF are
- Direct Sanger sequencing
- High-resolution melting analysis
- Pyrosequencing
- Real-time PCR

Note 5: If BRAF is positive and there is no mention of the mutated codon, or the mutated codon is not specified, code 4.

Note 6: If neoadjuvant therapy is given, record the assay from tumor specimens prior to neoadjuvant therapy.
- If neoadjuvant therapy is given and there are no BRAF results from pre-treatment specimens, report the findings from post-treatment specimens

Code	Description
0	Normal BRAF negative, BRAF wild type Negative for (somatic) mutations, no alterations, no (somatic) mutations identified, not present, not detected
1	Abnormal (mutated)/detected: BRAF V600E (c.1799T>A) mutation

Code	Description
2	Abnormal (mutated)/detected, but not BRAF V600E (c.1799T>A) mutation
3	Abnormal (mutated)/detected, *KIAA1549*:*BRAF* gene fusion
4	Abnormal (mutated) BRAF, NOS
7	Test ordered, results not in chart
8	Not applicable: Information not collected for this case (If this information is required by your standard setter, use of code 8 may result in an edit error.)
9	Not documented in medical record Cannot be determined by pathologist BRAF not assessed or unknown if assessed
<BLANK>	N/A - Diagnosis year is prior to 2024

3c1-3c4, 3e: Medulloblastoma

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C700-C729	9470-9472-9478, 9480, 9501-9504, 9508	All ages	0, 1, 3
C700-C729, C753	9362	All ages	0, 1, 3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Medulloblastoma, Toronto Staging is based on the presence or absence of mets based on the *Chang M* definition for metastases within the CNS at diagnosis and is collected in Pediatric Mets.

- Pediatric Primary Tumor is collected for surveillance purposes

3c1-3c4, 3e: Medulloblastoma

9623: Pediatric Primary Tumor

Coding Instructions and Codes

Note 1: Pediatric Primary Tumor for Medulloblastoma is coded **only for single tumors confined to the primary site** (see code 150) or a **single** tumor crossing the midline without extension to adjacent structures (see code 250).

- Code 999 if there are multiple tumors in the brain
- The presence of multiple tumors is recorded in Pediatric Mets

Note 2: Benign (/0) or Borderline (/1) tumors are **always coded to 050** regardless of size or extension to adjacent sites

Note 3: A midline shift is not the same thing as crossing the midline

- Documentation must state "crosses/crossing the midline"
- Code 150 if you have a single tumor confined to the primary site with a midline shift that is not extending into adjacent structures (see Note 4).

Note 4: Direct or contiguous extension to an adjacent site is collected in Pediatric Mets.

- If the only information available is extension to an adjacent site, code Pediatric Primary Tumor 999 and assign the appropriate Pediatric Mets code
- The following are collected in Pediatric Mets (see code 25 for all except circulating cells in CSF (code 15))
    - Adjacent connective/soft tissue
    - Adjacent muscle
    - Bone
    - Circulating cells in cerebral spinal fluid (CSF)
    - Major blood vessel(s)
    - Meninges (e.g., dura)
    - Multiple/multifocal tumors
    - Nerves (cranial, NOS)
    - Ventricular system

Code	Description	SS2018 T
050	Benign or borderline brain	B
150	All sites • Single tumor confined to the primary site with no invasion or seeding to other structures Confined to site of origin, NOS Localized, NOS	L
250	Single tumor confined to the primary site that crosses/crossing the midline • WITHOUT invasion of adjacent structures (see Note 4)	RE
800	No evidence of primary tumor	U
999	Unknown; extension not stated Multiple tumors (See Note 1) Single tumor with extension to an adjacent site (see Note 4) Primary tumor cannot be assessed Not documented in medical record Death Certificate Only	U

3c1-3c4, 3e: Medulloblastoma

9624: Pediatric Regional Nodes

- Not applicable (code 888)

9625: Pediatric Mets

Coding Instructions and Codes

Note 1: Use code 70 when the only information is "distant metastasis, NOS," and there is no documentation regarding the specific metastases.

- If there are specific metastasis documented that are not listed in codes 15, 25, or 35, or 45, assign code 45 for "other specified distant metastasis."

Note 2: Code 00 for benign (behavior /0) and borderline (behavior /1) tumors.

Note 3: The following adjacent structures/sites, by direct or contiguous extension, are coded to 35.

- Adjacent connective/soft tissue
- Adjacent muscle
- Bone
- Circulating cells in cerebral spinal fluid (CSF)
- Major blood vessel(s)
- Meninges (e.g.; dura)
- Multiple/multifocal tumors
- Nerves (cranial, NOS)
- Ventricular system

Note 4: Leptomeningeal metastases, also known as carcinomatous meningitis and meningeal carcinomatosis, refers to the spread of malignant cells through the CSF space. These cells can originate from primary CNS tumors (e.g., in the form of drop metastases), as well as from distant tumors that have metastasized via hematogenous spread (code 35).

Code	Description	SS2018 M
00	No visible disease on imaging (MRI brain and spine) beyond primary site of disease AND no tumor cells into the cerebrospinal fluid (CSF)	None
15	Tumor cells in the CSF • Circulating cells in cerebral spinal fluid (CSF)	D

Code	Description	SS2018 M
25	Intracranial spread beyond a single lesion All sites • Bone (skull) (see code 45 for other bone involvement) • Major blood vessel(s) • Meninges (e.g., dura) • Multiple/multifocal tumors • Nerves (cranial, NOS) • Tumor invades or encroaches upon ventricular system Brain tumors (C700, C710-C719) • Anterior cranial fossa • Brain stem • Cerebellum • Cerebrum (cerebral hemisphere) • Contralateral hemisphere • Hypothalamus • Middle cranial fossa • Pallium • Posterior cranial fossa • Suprasellar brain • Tapetum • Thalamus CNS tumors (C701, C709, C720-C729) • Adjacent connective tissue • Adjacent muscle • Brain for cranial nerve tumor(s) • Sphenoid and frontal sinuses(skull) Pineal Gland (C753) • Adjacent connective/soft tissue • Cavernous sinus • Infratentorial and central brain	D
35	Visible metastasis in spine OR Visible metastasis in cervicomedullary (junction) Metastasis within CNS and CSF pathways • Carcinomatous meningitis • Drop metastasis • Leptomeningeal metastases • Meningeal carcinomatosis	D

Code	Description	SS2018 M
45	Extra-neural metastasis All Sites • Blood • Bone (other than skull) (see code 25 for skull) • Bone marrow • Carcinomatosis • Distant lymph nodes, NOS • Further contiguous extension • Other specified metastasis Brain tumors (C700, C710-C719) • Nasal cavity • Nasopharynx • Other direct extension outside CNS • Posterior pharynx CNS tumors (C701, C709, C720-729) • Eye	D
70	Distant metastasis, NOS	D
99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only	None

4a: Neuroblastoma

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C000-C699, C739-C750, C754-C809	9490, 9500	All ages	3
C700-C729, C751-C753	9490, 9500	All ages	0, 1, 3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- Children's Oncology Group (Newly Diagnosed with Neuroblastoma (childrensoncologygroup.org))
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Neuroblastoma, there are two different staging systems collected.

- Toronto Staging uses the *International Neuroblastoma Risk Group Staging System (INRGSS)* and records the stage group only. This is a clinical evaluation only based on image defined risk factors. This information will be collected in 9611: Intl Neuroblastoma Risk Grp Stag Sys (INRGSS)
- The Children's Oncology Group (COG) use the *International Neuroblastoma Staging System (INSS),* which is based on surgical resection and is defined by stage group
    - Pediatric Primary Tumor, Pediatric Regional Nodes, and Pediatric Mets will be collected for surveillance purposes based on surgical resection and will derive the INSS Stage Group

4a: Neuroblastoma

9623: Pediatric Primary Tumor

Coding Instructions and Codes

Note 1: This field is based on surgical resection of the primary site only WITH or WITHOUT neoadjuvant therapy.

- Code 100 (localized tumor) OR 300 (regional tumor) if a procedure removes the entire tumor (i.e.; surgical resection, excisional biopsy)
- Code 200 (localized tumor) OR 400 (regional tumor) if a procedure does not remove the entire tumor (i.e.; incomplete, partial), or it's not clear if the entire tumor was removed
- Code 600 if the tumor cannot be surgically removed or the tumor crosses the spine (one side of the body to the other side)
- Code 999 if there was no surgical removal of the tumor, or there was a clinical work up only

Note 2: Use the following resources to determine if a tumor is localized, regional, or distant (further contiguous extension).

- EOD: Review the primary site-based schema
- Summary Stage: Review the primary site-based chapter

Code	Description	SS2018 T
100	Localized tumor confined to one side of the body and one area • Tumor completely surgically resected • Localized, NOS	L
200	Localized tumor confined to one side of the body, greater than one area • Incomplete/partial surgical resection done OR unknown if complete surgical resection done • Unable to resect tumor	L
300	Regional tumor confined to one side of the body and one area • Tumor completely surgically resected	RE
400	Regional tumor confined to one side of the body, greater than one area • Incomplete/partial surgical resection done OR unknown if complete surgical resection done • Unable to resect tumor	RE
600	Tumor starts in or crosses the vertical midline (spine) of the body Cannot be removed surgically	D
700	Further contiguous extension	D
800	No evidence of primary tumor	U
999	Unknown; extension not stated Primary tumor cannot be assessed Not documented in medical record Surgical resection not done/recommended Unknown if surgery done Clinical workup only Death Certificate Only	U

4a: Neuroblastoma

9624: Pediatric Regional Nodes

Coding Instructions and Codes

Note 1: This field is based on clinical and/or pathological information WITH or WITHOUT neoadjuvant therapy.

Note 2: Code only regional nodes, and nodes, NOS in this field. Distant nodes are coded in Pediatric Mets.

Note 3: Use the following resources to determine if involved lymph nodes are regional or distant.

- EOD: Review the primary site-based schema
- Summary Stage: Review the primary site-based chapter

Code	Description	SS2018 N
000	No regional lymph node involvement	NONE
100	Ipsilateral regional lymph nodes involved	RN
300	Contralateral or bilateral regional lymph nodes involved	RN
800	Regional lymph node(s), NOS Lymph node(s), NOS Unknown if ipsilateral, contralateral or bilateral	RN
888	Not applicable: Primary site C420-C424, C700-C729, C751-C753, C760-C809	NA
999	Unknown; regional lymph node(s) not stated Regional lymph node(s) cannot be assessed Not documented in medical record Death Certificate Only	U

4a: Neuroblastoma

9625: Pediatric Mets

Coding Instructions and Codes

Note: Use the following resources to determine if involved lymph nodes are distant or if involved organs are metastatic.

- EOD: Review the primary site-based schema
- Summary Stage: Review the primary site-based chapter

Code	Description	SS2018 M
00	No distant metastasis	None
10	Distant lymph node(s), NOS	D
20	Skin WITH or WITHOUT distant lymph nodes	D
30	Liver WITH or WITHOUT distant lymph nodes or skin	D
40	Bone marrow WITH or WITHOUT distant lymph nodes, skin, or liver	D
50	Bone WITH or WITHOUT distant lymph nodes, skin, liver, bone marrow	D
70	Other specified metastasis excluding skin, liver, bone marrow, bone Carcinomatosis Distant metastasis WITH or WITHOUT distant lymph node(s) Distant metastasis, NOS	D
99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only	None

4a: Neuroblastoma

9611: Intl Neuroblastoma Risk Grp Stag Sys (INRGSS)

Item Length: 1
NAACCR Item #: 9611
XML NAACCR ID: inrgss
Active years: TBD
Schema(s):

- 4a: Neuroblastoma

Description

International Neuroblastoma Risk Group Staging System (INRGSS) for Neuroblastoma is defined based on clinical work up and image-defined risk factors.

Rationale

This is part of the National Childhood Cancer Registry (NCCR) project to collect more specific information on pediatric patients. Registries part of the NCCR will start collection on specific pediatric data items with 2024+ diagnoses.

Definition

This staging system is able to be utilized for every single neuroblastoma tumor as it is based on the clinical work up and image-defined risk factors.

Additional Information:

- **Other names:** INRG

Coding Instructions and Codes

Possible sources of information: imaging

Note 1: This SSDI is effective for diagnosis years 2024+

- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: The INRGSS is a clinical based staging system and is able to be utilized for every single neuroblastoma tumor as it is based on the clinical work up and image-defined risk factors

- Primary source of information for this data item is imaging
- Do not code any staging information from results of surgical resection in this data item
    - *Note*: There is a different staging system available for patients who have surgery (International Neuroblastoma Staging System (INSS)) which is collected in Pediatric Primary Tumor, Pediatric Regional Nodes, and Pediatric Mets

Note 3: Image-Defined Risk Factors in Neuroblastic Tumors: Staging requires assessment of whether patients have none, or one or more of the image-defined risk factors (IDRF). These IDRF's are based on imaging prior to any surgical resection or other treatment.

- Ipsilateral tumor extension within two body compartments
    - Neck-chest, chest-abdomen, abdomen-pelvis
- Neck
    - Tumor encasing carotid and/or vertebral artery and/or internal jugular vein
    - Tumor extending to base of skull

  - Tumor compressing the trachea
- Cervico-thoracic junction
  - Tumor encasing brachial plexus roots
  - Tumor encasing subclavian vessels and/or vertebral and/or carotid artery
  - Tumor compressing the trachea
- Thorax
  - Tumor encasing the aorta and/or major branches
  - Tumor compressing the trachea and/or principal bronchi
  - Lower mediastinal tumor, infiltrating the costovertebral junction between T9 and T12
- Thoraco-abdominal
  - Tumor encasing the aorta and/or vena cava
- Abdomen, pelvis
  - Tumor infiltrating the porta hepatis and/or the hepatoduodenal ligament
  - Tumor encasing branches of the superior mesenteric artery at the mesenteric root
  - Tumor encasing the origin of the coeliac axis, and/or of the superior mesenteric artery
  - Tumor invading one or both renal pedicles
  - Tumor encasing the aorta and/or vena cava
  - Tumor encasing the iliac vessels
  - Pelvic tumor crossing sciatic notch
- Intraspinal tumor extension whatever the location provided that
  - More than one third of the spinal canal in the axial plane is invaded and/or the perimedullary leptomeningeal spaces are not visible and/or the spinal cord signal is abnormal
- Infiltration of adjacent organs/structures
  - Pericardium, diaphragm, kidney, liver, duodeno-pancreatic block, and mesentery

Note 4: Ascites and/or a pleural effusion, even with malignant cells, do not constitute metastatic disease unless they are remote from the body compartment of the primary tumor.

Note 5: Regional lymph node involvement does not factor into staging

- Code 3 for non-regional lymph node involvement (distant lymph nodes)

Code	Description
1	Stage L1 • Localized tumor that does not involve any vital structures • Tumor confined within one body compartment (i.e., neck, chest, abdomen, or pelvis) • No evidence of image-defined risk factors (IDRF's) • Intraspinal tumor extension that does not fulfil the criteria for an IDRF is consistent with stage (L1)
2	Stage L2 • Locoregional tumor with evidence of image-defined risk factors (IDRF's) • Tumor ipsilaterally contiguous within body compartments (i.e., a left sided abdominal tumor with left-sided lung, bone, or pleura involvement) ○ Non-contiguous disease would be coded as M (e.g., left sided abdominal tumor with right-sided lung, bone, or pleura)
3	Stage M • Distant metastatic disease (see Stage MS for patients less than 18 months) • Noncontiguous disease • Distant lymph node involvement

Code	Description
4	Stage MS • For patients less than 18 months only (547 days) metastatic disease confined to o Bone marrow ▪ MIBG scintigraphy must be negative in bone and bone marrow o Skin o Liver
8	Not applicable: Information not collected for this case (If this information is required by your standard setter, use of code 8 may result in an edit error)
9	Not documented in medical record International Neuroblastoma Risk Group Staging System not assessed Death certificate only
<BLANK>	N/A - Diagnosis year is prior to 2024

4a: Neuroblastoma

9614: n-MYC Amplification

Item Length: 1
NAACCR Item #: 9614
XML NAACCR ID: nMycAmplification
Active years: TBD
Pediatric Schema(s):

- 4a: Neuroblastoma

Description

n-MYC Amplification is a gene that normally regulates cell growth. Studies have shown that n-MYC amplification is an indicator of poor prognosis and increased risk of unfavorable outcomes in patients with neuroblastoma.

Rationale

This is part of the National Childhood Cancer Registry (NCCR) project to collect more specific information on pediatric patients. Registries part of the NCCR will start collection on specific pediatric data items with 2024+ diagnoses.

Definition

This field indicates the n-MYC status of a neuroblastoma tumor after a pathologic specimen is obtained.

Coding Instructions and Codes

Note 1: This SSDI is effective for diagnosis years 2024+

- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: n-MYC Amplification is a gene that normally regulates cell growth. Studies have shown that n-MYC amplification is an indicator of poor prognosis and increased risk of unfavorable outcomes in patients with neuroblastoma.

- Primary sources of information: molecular pathology report (may be addendum to original pathology report), CAP Protocol

Note 3: Physician statement of n-MYC can be used to code this data item when no other information is available.

Code	Description
0	Not amplified/negative
1	Amplified/positive
2	Gain
7	Test ordered, results not in chart
8	Not applicable: Information not collected for this case (If this information is required by your standard setter, use of code 8 may result in an edit error)
9	Not documented in medical record Cannot be determined by pathologist Not applicable (secondary to previous chemotherapy) n-MYC not assessed or unknown if assessed
<BLANK>	N/A - Diagnosis year is prior to 2024

4a: Neuroblastoma

9610: Intl Neuroblastoma Path Prog Class (INPC)

Item Length: 1
NAACCR Item #: 9610
Alternate name: International Neuroblastoma Pathology Prognostic Classification (INPC), Shimada Classification, Unfavorable histology, or Favorable histology
XML NAACCR ID: inpc
Active years: TBD
Pediatric Schema(s):

- 4a: Neuroblastoma

Description

The International Neuroblastoma Pathology Prognostic Classification (INPC) categorizes neuroblastomas as favorable or unfavorable histologies based on the following factors: age, neuroblastic maturation, Schwannian stromal content, Mitosis-kayorrhexis index (MKI), and degree of differentiation (grade).

Rationale

This is part of the National Childhood Cancer Registry (NCCR) project to collect more specific information on pediatric patients. Registries part of the NCCR will start collection on specific pediatric data items with 2024+ diagnoses.

Coding Instructions and Codes

Note 1: This SSDI is effective for diagnosis years 2024+

- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: The International Neuroblastoma Pathology Prognostic Classification (INPC) (unfavorable versus favorable histology) is a biologically relevant classification based on morphologic features of neuroblastic tumors which has prognostic indications. It is based on the following criteria

- Age
- Neuroblastic maturation
- Schwannian stromal content
- Mitosis-kayorrhexis index (MKI)
- Degree of differentiation (grade)

Note 3: The INPC results are to be coded based on either biopsy or surgical resection (without neoadjuvant therapy). If the only results are from a post neoadjuvant surgical resection, code 9.

Note 4: Physician statement of unfavorable vs favorable histology can be used to code this data item when no other information is available, provided there is information that the results are from a biopsy or surgical resection without neoadjuvant therapy.

Code	Description
0	Unfavorable
1	Favorable
7	Test ordered, results not in chart
8	Not applicable: Information not collected for this case (If this information is required by your standard setter, use of code 8 may result in an edit error)

Code	Description
9	Not documented in medical record Cannot be determined by pathologist International Neuroblastoma Pathology Prognostic Classification (INPC) not assessed or unknown if assessed INPC assessed only at post neoadjuvant surgical resection
<BLANK>	N/A - Diagnosis year is prior to 2024

5: Retinoblastoma

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C690-C699	9510-9514	All ages	3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Retinoblastoma, Toronto Staging is based on the *International Retinoblastoma Staging System (IRSS)* and records the stage group only.

- Pediatric Primary Tumor, Pediatric Regional Nodes, and Pediatric Mets will be collected for surveillance purposes and will derive the IRSS Stage Group

5: Retinoblastoma

9623: Pediatric Primary Tumor

Coding Instructions and Codes

Note 1: If there are bilateral retinoblastomas (both eyes involved), record the stage of the eye with the more advanced/higher stage in this data item.

- Code the eye with the lesser/lower stage in the data item IRSS Stage for Eye-2 [NAACCR Data Item #9627]

Note 2: Pathological staging information from an enucleation always takes precedence over clinical staging, except in cases with neoadjuvant treatment where clinical disease is as extensive as or more extensive than disease at surgery.

Code	Description	SS2018 T
100	Intraocular tumor(s) WITHOUT any • Local invasion • Focal choroidal invasion • Pre- or intralaminar involvement of the optic nerve head Tumor confined to retina, NOS Localized, NOS	L
200	Intraocular tumor(s) WITH local invasion, NOS • Choroid (concomitant focal invasion) • Pre- or intralaminar involvement of optic nerve head • Retinal detachment • Schlemm's canal • Stromal invasion iris • Subretinal seeding • Trabecular meshwork • Vitreous seeding	L
300	Advanced intraocular tumor(s) WITH significant local invasion • Anterior chamber • Aseptic orbital cellulitis • Buphthalmos • Choroid (multiple foci, focal, full-thickness involvement) • Ciliary body • Emissary channels • Hyphema AND/OR massive vitreous hemorrhage • Iris • Lens • Pars plana • Phthisis or pre-phthisis bulbi • Raised intraocular pressure with neovascularization • Retrolaminar invasion of optic nerve head • Sclera • Zonules	L

Code	Description	SS2018 T
400	Evidence of extraocular tumor • Tumor at transected end of optic nerve • Tumor in the meningeal spaces around optic nerve Full-thickness invasion of sclera WITH invasion of • Adjacent adipose tissue • Bone • Conjunctiva • Episcleral • Extraocular muscle • Eyelids	RE
800	No evidence of primary tumor	U
999	Unknown; extension not stated Primary tumor cannot be assessed Not documented in medical record Death Certificate Only	U

5: Retinoblastoma

9624: Pediatric Regional Nodes

Coding Instructions and Codes

Note 1: Code only regional nodes and nodes, NOS, in this field. Distant nodes are coded in Pediatric Mets.

Note 2: Code 800 if regional lymph nodes are involved, but there is no indication which ones are involved

Code	Description	SS2018 N
000	No regional lymph node involvement	None
300	Cervical, NOS Mandibular, NOS • Submandibular (submaxillary) Parotid, NOS • Infra-auricular • Preauricular	RN
800	Regional lymph node(s), NOS Lymph nodes, NOS	RN
999	Unknown; regional lymph node(s) not stated Regional lymph node(s) cannot be assessed Not documented in medical record Death Certificate Only	U

5: Retinoblastoma

9625: Pediatric Mets

Coding Instructions and Codes

Note: Code 50 does not include "trilateral retinoblastomas." The presence of "trilateral retinoblastomas" is coded in the data item Heritable Trait [NAACCR Data Item #3856].

Code	Description	SS2018 M
00	No distant metastasis	None
10	Distant lymph node(s), NOS	D
30	Distant metastasis to any organ EXCEPT CNS Carcinomatosis Code 10 + 30	D
50	CNS parenchyma Cerebrospinal fluid Any combination of codes 10, 30 and 50	D
70	Distant metastasis, NOS	D
99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only	None

5: Retinoblastoma

9627: IRSS Stage for Eye-2

Item Length: 1
Provisional NAACCR Item #: 9627
Provisional XML Parent-NAACCR ID: irssStageForEye2
Active years: 2024+
Pediatric Schema(s):

- Retinoblastoma

Description

Bilateral retinoblastoma is abstracted as a single primary regardless of timing, but there is no effective way to measure the different extent of diseases or stages for each eye beyond text. Currently, abstractors are required to enter the information of the most advanced eye in the staging data fields, losing all measurable data on the stage the less advanced eye.

Rationale

Even though staging information for only one retinoblastoma tumor can currently be captured in data form, abstractors are still required to put treatment information for bilaterally affected eyes into the treatment fields of the abstract. The ability to capture measurable staging data of the contralateral eye will provide substantiation of the treatment captured for both eyes in the abstract.

Definition

This field allows the abstractor to enter the individual stages for each eye in cases of bilateral retinoblastoma at the time of diagnosis.

Provisional Coding Instructions and Codes

Possible sources of information: Imaging, Physician notes

Note 1: This SSDI is effective for diagnosis years 2024+

- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: This data item records the stage of the second eye when there are bilateral retinoblastomas.

- Code 7 if only one eye is involved (unilateral retinoblastoma)
- Extension of one eye (unilateral retinoblastoma) is coded in Pediatric Primary Tumor

Note 3: If there are bilateral retinoblastomas, record the Stage Group of the lesser/lower stage in this data item (more advanced/higher stage is recorded in Pediatric Primary Tumor).

Note 4: Do not record a stage (Stage 0-IV) in this data item if only one eye is involved (See Note 2).

Code	Description
0	Stage 0 • Tumor confined to the globe • Enucleation has not been performed • Patient treated conservatively with either focal therapies or chemotherapy

Code	Description
1	Stage I • Enucleation with negative margins (R0) • Completely resected histologically
2	Stage II • Enucleation with positive/microscopic residual tumor

3	Stage III
	• Regional extension, involvement of
	  ○ Orbit
	  ○ Preauricular extension
	  ○ Cervical lymph node involvement

4	Stage IV • Distant metastatic disease
7	Not applicable: Only one eye is involved, and staging collected in Pediatric Primary Tumor, Pediatric Regional Nodes and Pediatric Mets • Unilateral retinoblastoma
8	Not applicable: Information not collected for this case (If this information is required by your standard setter, use of code 8 may result in an edit error.)
9	Not documented in medical record International Retinoblastoma Staging System (IRSS) not assessed or unknown if assessed
<BLANK>	N/A - Diagnosis year is prior to 2024

6a1-6a4, 6c: Renal Tumors

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C649	8000-8005	00-39	3
C649, C659	8964-8967	00-39	3
C649, C659	8959, 8960	All ages	3
C649	8963, 9364	All ages	3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Renal tumors, Toronto Staging is based on the *Wilms Tumor Staging System* and records the stage group only.

- Pediatric Primary Tumor, Pediatric Regional Nodes, and Pediatric Mets will be collected for surveillance purposes and will derive the Wilms Tumor Stage Group

Note 3: This staging system is not the same as the National Wilms Tumor Study Group (NWTS)

- Do not record stage information from the National Wilms Tumor Study Group (NWTS) in these data items

6a1-6a4, 6c: Renal Tumors

9623: Pediatric Primary Tumor

Coding Instructions and Codes

Note 1: Primary tumor evaluation is based on a surgical resection of the primary site only.

- Code 999 if there is no surgical resection of the primary site
- Do not use imaging findings to code this data item

Note 2: Codes are based on whether patient had neoadjuvant chemotherapy or surgery first

- If patient did not receive chemotherapy (or unknown if patient received chemotherapy) prior to surgery, see codes 100, 200, 300
- If patient did receive chemotherapy prior to surgery, see codes 110, 210, 310

Note 3: If there is bilateral disease, code the most advanced stage at diagnosis.

- Code 4 for Laterality [NAACCR # 410] data item

Code	Description	SS2018 T

100	No chemotherapy AND no biopsy prior to surgery (see code 300 if biopsy done prior to surgical resection)
	• Tumor limited to kidney and completely resected
	  ○ Renal capsule intact, not penetrated by tumor
	  ○ No tumor invasion of veins or lymphatics of renal sinus
	• Confined (limited) to the kidney, NOS
	• Localized, NOS
	• WITH NEGATIVE or UNKNOWN margins (see code 300 if positive margins)
		L

110	Chemotherapy prior to surgery
	• Tumor limited to kidney and completely resected
	  ○ Renal capsule may be infiltrated by tumor, but tumor does not reach the outer surface
	  ○ Tumor may protrude or bulge into the pelvic system or ureter, but does not infiltrate
	  ○ Vessels of renal sinus not involved
		L

200	No chemotherapy AND no biopsy prior to surgery (see code 300 if biopsy done prior to surgical resection)
	• Tumor extends beyond kidney but completely resected
	  ○ Tumor penetrates renal capsule
	  ○ Tumor in lymphatics or veins of renal sinus
	  ○ Tumor in renal vein with margin not involved
	• WITH NEGATIVE or UNKNOWN margins (see code 300 if positive margins)
		RE

210	Chemotherapy prior to surgery
	• Tumor extends beyond kidney but completely resected
	  ○ Tumor penetrates renal capsule into perirenal fat
	  ○ Tumor infiltrates the renal sinus and/or invades blood and lymphatic vessels outside renal parenchyma but is completely resected
	  ○ Tumor infiltrates adjacent organs or vena cava but is completely resected
		RE

Code	Description	SS2018 T

300	No chemotherapy prior to surgery
	• Residual tumor confined to abdomen
	  ○ Peritoneal contamination or tumor implant
	  ○ Tumor spillage (rupture) of any degree occurring before or during surgery
	  ○ Gross residual tumor in abdomen
	• WITH POSITIVE margins (includes localized tumor with positive margins)
	OR
	• Biopsy of tumor (including fine-needle aspiration) prior to surgical resection of kidney
		RE

310	Chemotherapy prior to surgery
	• Incomplete resection of the tumor (gross or microscopic extension beyond the resection margins)
	  ○ Necrotic tumor or chemotherapy-induced changes
	  ○ Tumor rupture before or intraoperatively
	  ○ Tumor has penetrated the peritoneal surface
	  ○ Tumor thrombi present at resection margins
	  ○ Surgical biopsy prior to resection (does not include needle biopsy)
		RE

800	No evidence of primary tumor	U
999	Unknown; extension not stated Primary tumor cannot be assessed Not documented in medical record No surgical resection of the primary site Death certificate only	U

6a1-6a4, 6c: Renal Tumors

9624: Pediatric Regional Nodes

Coding Instructions and Codes

Note 1: Regional lymph node evaluation is based on microscopic evaluation (FNA, biopsy, sentinel lymph node biopsy, lymph node dissection) only.

- Code 999 if there is no microscopic evaluation of regional lymph nodes
- Do not use imaging findings to code this data item

Note 2: Code only regional nodes and nodes, NOS, in this field. Distant nodes are coded in Pediatric Mets.

Note 3: Regional lymph nodes are defined as those in the vicinity of the primary tumor.

Code	Description	SS2018 N
000	No regional lymph node involvement	NONE
300	Aortic, NOS • Lateral (lumbar) • Para-aortic • Periaortic • Preaortic • Retroaortic Caval, NOS • Interaortocaval • Paracaval • Pericaval • Precaval • Retrocaval Renal hilar Retroperitoneal, NOS	RN
800	Regional lymph node(s), NOS Lymph node(s), NOS	RN
999	Unknown; regional lymph node(s) not stated Regional lymph node(s) cannot be assessed Not documented in medical record No microscopic evaluation of regional lymph nodes Death Certificate Only	U

6a1-6a4, 6c: Renal Tumors

9625: Pediatric Mets

Coding Instructions and Codes

Note: The presence of mets can be based on imaging and/or microscopic evaluation.

Code	Description	SS2018 M
00	No distant metastasis	NONE
10	Distant lymph node(s), NOS	D

70	Extension to
	
	• Adrenal gland
	  ○ Ipsilateral, noncontiguous
	  ○ Contralateral
	• Contralateral kidney
	• Contralateral ureter
	• Liver
	• Spleen
	
	Carcinomatosis
	
	Distant metastasis WITH or WITHOUT distant lymph node(s)
	
	Distant metastasis, NOS
		D

99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only	None

6a1-6a4, 6c-Renal Tumors

3801: Chromosome 1p: Loss of Heterozygosity

Item Length: 1
NAACCR Item #: 3801
XML NAACCR ID: chromosome1pLossHeterozygosity
Active years: 2024+
Pediatric Schema(s):

- 6a1: Renal Tumors: Nephroblastoma
- 6a2: Renal Tumors: Rhabdoid Renal Tumor
- 6a3: Renal Tumors: Kidney Sarcomas
- 6a4: Renal Tumors: Ewing Sarcoma of Kidney
- 6c: Renal Tumors: Unspecified

Description

Chromosome 1p: Loss of Heterozygosity (LOH) refers to the loss of genetic material normally found on the short arm of one of the patient's two copies of chromosome 1. Occurs in approximately 5% of favorable histology (non-anaplastic) Wilm's tumor (FHWT) cells and has been shown to be associated with inferior relapse-free survival (RFS) and overall survival (OS) in patients with FH Wilm's tumor. This testing is commonly done in conjunction with Chromosome 1q: Loss of Heterozygosity (NAACCR ID: 9601) and Chromosome 16q: Loss of Heterozygosity (NAACCR ID: 9600).

Rationale

This is part of the National Childhood Cancer Registry (NCCR) project to collect more specific information on pediatric patients. Registries part of the NCCR will start collection on specific pediatric data items with 2024+ diagnoses.

Coding Instructions and Codes

Note 1: This SSDI is effective for diagnosis years 2024+.

- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: Chromosome 1p: Loss of Heterozygosity (LOH) refers to the loss of genetic material normally found on the short arm of one of the patient's two copies of chromosome 1. Occurs in approximately 5% of favorable histology (non-anaplastic) Wilm's tumor (FHWT) cells and has been shown to be associated with inferior relapse-free survival (RFS) and overall survival (OS) in patients with FH Wilm's tumor. This testing is commonly done in conjunction with Chromosome 1q: Loss of Heterozygosity (NAACCR ID: 9601) and Chromosome 16q: Loss of Heterozygosity (NAACCR ID: 9600).

- This is a special molecular diagnostic test performed on tumor tissue to identify loss of genetic material normally found on the short arm of one of the patient's two copies of chromosome 1. A normal cell will contain two complete copies of each chromosome, one from each parent, and this normal state is termed heterozygous. Loss of heterozygosity (LOH) is an abnormal state reflecting loss of the whole arm of chromosome 1p following a chromosomal translocation event
- Primary sources of information: molecular pathology report (may be addendum to original pathology report)

Note 3: Physician statement of Chromosome 1p deletion/LOH can be used to code this data item.

Note 4: Other terms for LOH include whole arm loss, gene deletion and allelic loss.

Code	Description
0	Chromosome 1p deletion/LOH not identified/not present/negative
1	Chromosome 1p deletion/LOH identified/present/positive

Code	Description
7	Test ordered, results not in chart
8	Not applicable: Information not collected for this case (If this information is required by your standard setter, use of code 88 may result in an edit error.)
9	Not documented in medical record Cannot be determined by pathologist Chromosome 1p deletion/LOH not assessed or unknown if assessed
<BLANK>	N/A - Diagnosis year is prior to 2024

6a1-6a4, 6c-Renal Tumors

9600: Chromosome 16q: Loss of Heterozygosity

Item Length: 1
NAACCR Item #: 9600
XML NAACCR ID: chromosome16qLossHeterozygosity
Active years: 2024+
Pediatric Schema(s):

- 6a1: Renal Tumors: Nephroblastoma
- 6a2: Renal Tumors: Rhabdoid Renal Tumor
- 6a3: Renal Tumors: Kidney Sarcomas
- 6a4: Renal Tumors: Ewing Sarcoma of Kidney
- 6c: Renal Tumors: Unspecified

Description

Chromosome 16q: Loss of Heterozygosity (LOH) refers to the loss of genetic material normally found on the short arm of one of the patient's two copies of chromosome 16. Occurs in approximately 5% of favorable (non-anaplastic) histology Wilm's tumor (FHWT) cells and has been shown to be associated with inferior relapse-free survival (RFS) and overall survival (OS) in patients with FH Wilm's tumor. This testing is commonly done in conjunction with Chromosome 1p: Loss of Heterozygosity (NAACCR ID: 3801) and Chromosome 1q: Loss of Heterozygosity (NAACCR ID: 9601)

Rationale

This is part of the National Childhood Cancer Registry (NCCR) project to collect more specific information on pediatric patients. Registries part of the NCCR will start collection on specific pediatric data items with 2024+ diagnoses.

Coding Instructions and Codes

Note 1: This SSDI is effective for diagnosis years 2024+.

- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: Chromosome 16q: Loss of Heterozygosity (LOH) refers to the loss of genetic material normally found on the short arm of one of the patient's two copies of chromosome 16. Occurs in approximately 5% of favorable (non-anaplastic) histology Wilm's tumor (FHWT) cells and has been shown to be associated with inferior relapse-free survival (RFS) and overall survival (OS) in patients with FH Wilm's tumor. This testing is commonly done in conjunction with Chromosome 1p: Loss of Heterozygosity (NAACCR ID: 3801) and Chromosome 1q: Loss of Heterozygosity (NAACCR ID: 9601)

- This is a special molecular diagnostic test performed on tumor tissue to identify loss of genetic material found on the long arm of one of the patient's two copies of chromosome 16. A normal cell will contain two complete copies of each chromosome, one from each parent, and this normal state is termed heterozygous. Loss of heterozygosity (LOH) is an abnormal state reflecting loss of the whole arm of chromosome 16q following a chromosomal translocation event
- Primary sources of information: molecular pathology report (may be addendum to original pathology report)

Note 3: Physician statement of Chromosome 16q deletion/LOH can be used to code this data item.

Note 4: Other terms for LOH include whole arm loss, gene deletion and allelic loss.

Table begins on next page

Code	Description
0	Chromosome 16q deletion/LOH not identified/not present/negative
1	Chromosome 16q deletion/LOH present/positive
7	Test ordered, results not in chart
8	Not applicable: Information not collected for this case (If this item is required by your standard setter, use of code 8 will result in an edit error.)
9	Not documented in medical record Cannot be determined by pathologist Chromosome 16q: LOH not assessed or unknown if assessed
<BLANK>	N/A - Diagnosis year is prior to 2024

6a1-6a4, 6c-Renal Tumors

9601: Chromosome 1q Status

Item Length: 1
NAACCR Item #: 9601
XML NAACCR ID: chromosome1qStatus
Active years: 2024+
Pediatric Schema(s):

- 6a1: Renal Tumors: Nephroblastoma
- 6a2: Renal Tumors: Rhabdoid Renal Tumor
- 6a3: Renal Tumors: Kidney Sarcomas
- 6a4: Renal Tumors: Ewing Sarcoma of Kidney
- 6c: Renal Tumors: Unspecified

Description

Gain of chromosome 1q is one of the most common cytogenetic findings in Wilms tumor, occurring approximately 30% of tumors. It is associated with a poorer relapse-free survival (RFS) and overall survival (OS) in patients with favorable (non-anaplastic) histology Wilm's tumor (FHWT). This testing is commonly done in conjunction with Chromosome 1p: Loss of Heterozygosity (see NAACCR ID: 3801) and Chromosome 16q: Loss of Heterozygosity (see NAACCR ID: 9600).

Rationale

This is part of the National Childhood Cancer Registry (NCCR) project to collect more specific information on pediatric patients. Registries part of the NCCR will start collection on specific pediatric data items with 2024+ diagnoses.

Coding Instructions and Codes

Note 1: This SSDI is effective for diagnosis years 2024+.

- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: Gain of chromosome 1q is one of the most common cytogenetic findings in Wilms tumor, occurring approximately 30% of tumors. It is associated with a poorer relapse-free survival (RFS) and overall survival (OS) in patients with favorable (non-anaplastic) histology Wilm's tumor (FHWT). This testing is commonly done in conjunction with Chromosome 1p: Loss of Heterozygosity (see NAACCR ID: 3801) and Chromosome 16q: Loss of Heterozygosity (see NAACCR ID: 9600).

- This is a special molecular diagnostic test performed on tumor tissue to identify gain of genetic material normally found on the short arm of one of the patient's two copies of chromosome 1. A normal cell will contain two complete copies of each chromosome, one from each parent, and this normal state is termed heterozygous. Gain of heterozygosity (GOH) is an abnormal state reflecting gain of the whole arm of chromosome 1q following a chromosomal translocation event
- Primary sources of information: molecular pathology report (may be addendum to original pathology report)

Note 3: Physician statement of Chromosome 1q gain can be used to code this data item.

Code	Description
0	Chromosome 1q gain/GOH not identified/not present/negative
1	Chromosome 1q gain/GOH identified/present/positive
7	Test ordered, results not in chart
8	Not applicable: Information not collected for this case (If this information is required by your standard setter, use of code 88 may result in an edit error.)

Code	Description
9	Not documented in medical record Cannot be determined by pathologist Chromosome 1q gain/LOH not assessed or unknown if assessed
<BLANK>	N/A - Diagnosis year is prior to 2024

6a4-Renal Tumors: Ewing Sarcoma of Kidney

9608: EWSR1-FLI1 fusion

Item Length: 1
NAACCR Item #: 9608
XML NAACCR ID: ewsr1Fli1Fusion
Active years: 2024+
Pediatric Schema(s):

- 6a4: Renal Tumors: Ewing Sarcoma of Kidney
- 8c: Malignant Bone Tumors: Ewing Sarcoma

Description

EWS-FLI1 fusion occurs in about 90% of Ewing Sarcomas and functions as both a pioneering transcription factor and potent oncogene.

Rationale

This is part of the National Childhood Cancer Registry (NCCR) project to collect more specific information on pediatric patients. Registries part of the NCCR will start collection on specific pediatric data items with 2024+ diagnoses.

Coding Instructions and Codes

Note 1: This SSDI is effective for diagnosis years 2024+ .

- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: EWS-FLI1 fusion occurs in about 90% of Ewing Sarcomas and functions as both a pioneering transcription factor and potent oncogene.

- Primary sources of information: molecular pathology report (may be addendum to original pathology report), CAP protocol

Note 3: Physician statement of the EWSR1-FLI1 fusion can be used to code this data item.

Code	Description
0	No gene rearrangements (fusions) identified
1	EWSR1-FLI1 gene rearrangement (fusion) present
2	EWSR1-ERG gene rearrangement (fusion) present
3	Other EWSR1 gene rearrangement (fusion) present
4	EWSR1 rearrangement present, fusion partner not known
5	Non-EWSR1 variant translocation present
7	Test ordered, results not in chart
8	Not applicable: Information not collected for this case (If this information is required by your standard setter, use of code 8 may result in an edit error.)
9	Not documented in medical record Cannot be determined by pathologist EWSR1 gene arrangements not assessed or unknown if assessed
<BLANK>	N/A - Diagnosis year is prior to 2024

7a: Hepatoblastoma

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C220	8970	All ages	3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- Children's Oncology Group (Newly Diagnosed with Hepatoblastoma or Hepatocellular Carcinoma (childrensoncologygroup.org)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Hepatoblastoma tumors, there are three different staging systems collected

- Toronto Staging is based on the absence or presence of mets and is collected in Pediatric Mets
- The Children's Oncology Group (COG) for liver staging is used.
    - Per this staging system, anything involved outside the liver is Stage IV. This is different than other staging used in the US that has adjacent organs not being a Stage IV
    - Pediatric Primary Tumor, Pediatric Regional Nodes and Pediatric Mets are used to derive the stage group
- Pretext is collected as a SSDI and is based on clinical staging only

7a: Hepatoblastoma

9623: Pediatric Primary Tumor

Coding Instructions and Codes

Note 1: Evaluation of primary tumor is based on several factors

- Surgically resected WITH or WITHOUT neoadjuvant therapy and tumor(s) confined to the liver with **negative/unknown** margins
  - One lobe involved (see code 150)
  - More than one lobe involved (see code 250)
- Surgically resected WITH or WITHOUT neoadjuvant therapy and tumor(s) confined to the liver with **positive** margins (codes 175 and 275)
  - One lobe involved (see code 175)
  - More than one lobe involved (see code 275)
- Partial surgical resection WITH or WITHOUT neoadjuvant therapy, or unresectable and tumor(s) confined to the liver (code 350)

Note 2: See Pediatric Mets for involvement of adjacent structures, along with metastatic disease

Code	Description	SS2018 T
150	PATHOLOGICAL ASSESSMENT ONLY Tumor confined to the liver WITH or WITHOUT vascular invasion and NEGATIVE/UNKNOWN surgical margins • Complete resection of the tumor(s) • Single lesion (one lobe) • Multiple (satellite) nodules/tumors confined to one lobe • Confined to liver, NOS • Localized, NOS	L
175	Code 150 with positive surgical margins	L
250	PATHOLOGICAL ASSESSMENT ONLY Tumor confined to the liver WITH or WITHOUT vascular invasion • Complete resection of the tumor(s) • More than one lobe involved by contiguous growth (single lesion) • Multiple (satellite) nodules/tumors in more than one lobe of liver or on surface of parenchyma	RE
275	Code 250 with positive surgical margins	RE
350	Tumor confined to the liver WITH or WITHOUT vascular invasion • Incomplete resection done or no surgical resection performed • Determined to be unresectable • Presses onto vital tissues in the liver	RE
800	No evidence of primary tumor	U

Code	Description	SS2018 T
999	Unknown, extension not stated Primary tumor cannot be assessed Not documented in medical record Death Certificate Only	U

7a: Hepatoblastoma

9624: Pediatric Regional Nodes

Coding Instructions and Codes

Note 1: Code only regional nodes and nodes, NOS, in this field. Distant nodes are coded in Pediatric Mets.

Note 2: Regional lymph nodes are defined as those in the vicinity of the primary tumor.

Code	Description	SS2018 N
000	No regional lymph node involvement	NONE
300	Caval Hepatic, NOS • Hepatic artery • Hepatic pedicle • Inferior vena cava • Porta hepatis (portal) (hilar) [in hilus of liver] Hepatoduodenal ligament Periportal Portal vein	RN
700	Inferior phrenic nodes	D
800	Regional lymph node(s), NOS Lymph node(s), NOS	RN
999	Unknown; regional lymph node(s) not stated Regional lymph node(s) cannot be assessed Not documented in medical record Death Certificate Only	U

7a: Hepatoblastoma

9625: Pediatric Mets

Coding Instructions and Codes

Note: This field includes the involvement of adjacent structures to the liver.

Code	Description	SS2018 M
00	No distant metastasis	None
10	Diaphragm Extrahepatic bile duct(s) Extrahepatic blood vessel(s) • Hepatic artery • Portal vein • Vena cava Gallbladder Ligament(s) • Coronary • Falciform • Hepatoduodenal • Hepatogastric • Round (of liver) • Triangular Omentum (lesser and NOS) (see code 20 for greater omentum) Peritoneum, NOS • Parietal • Visceral	RE
20	Greater omentum (see code 10 for lesser omentum and omentum, NOS) Pancreas Pleura Stomach Further contiguous extension	D
30	Distant lymph node(s) • Aortic (para-aortic, periaortic) • Cardiac • Coronary artery • Diaphragmatic, NOS • Lateral (aortic) (lumbar) • Pericardial (pericardiac) • Peripancreatic (near head of pancreas only) • Posterior mediastinal (tracheoesophageal) including juxtaphrenic nodes • Renal artery • Retroperitoneal, NOS Distant lymph node(s), NOS	D
40	Lungs	D

Code	Description	SS2018 M
70	Carcinomatosis Distant metastasis WITH or WITHOUT distant lymph node(s) Other specified metastasis Distant metastasis, NOS	D
99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only	None

7a: Hepatoblastoma

9626: Pretext Clinical Staging

Item Length: 1
Provisional XML Parent-NAACCR ID: pretextClinicalStaging
Active years: 2024+
Pediatric Schema(s): Liver

Description

PRETEXT stands for PRE-Treatment Extent of tumor. This field describes the extent of involvement within the four lobes of the liver at time of a pediatric liver tumor diagnosis. It is based off clinical imaging and was originally designed to standardize imaging evaluation and risk stratification of hepatoblastoma before neoadjuvant chemotherapy or tumor resection.

Rationale

After initially being created by the International Childhood Liver Tumours Strategy Group (SIOPEL) in 1990, PRETEXT was introduced for use within the United States in 2014 by the Children's Oncology Group (COG) once radiographic imaging became more sophisticated and exploratory surgery at diagnosis was no longer advisable. It is used as a central component of risk stratification schemes that define treatment of hepatoblastoma.

Provisional Coding Instructions and Codes

Possible sources of information: Imaging

Note 1: This SSDI is effective for diagnosis years 2024+

- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: This SSDI is based on imaging findings only. Do not code any findings from surgical resection in this data item. Evaluation must also be done prior to any neoadjuvant therapy.

Note 3: For Pretext Staging, the 8 segments of the liver are grouped into sections

- Caudate liver: Segment I
  - *Note:* Involvement of the caudate liver is at minimum Stage 2
- Left lateral section: Segments II & III
- Left medial section: Segments IVA & IVB
- Right anterior section: Segments V & VIII
- Right posterior section: Segments VI & VII

Code	Description
1	One section involved; three adjoining sections are tumor free Stage I, Pretext 1
2	One or two sections involved; two adjoining sections are tumor free Stage 2, Pretext 2

Code	Description
3	Two or three sections involved; one adjoining section is tumor free Stage 3, Pretext 3
4	Four sections involved Stage 4, Pretext 4
8	Not applicable: Information not collected for this case (If this information is required by your standard setter, use of code 8 may result in an edit error.)
9	Not documented in medical record Cannot be determined by pathologist PRETEXT not assessed or unknown if assessed
<BLANK>	N/A- Diagnosis year is prior to 2024

8a-8e: Malignant Bone Tumors

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C400-C419, C760-C768, C809	9180-9187, 9191-9195, 9200, 9210, 9220-9221, 9230, 9240-9243, 9260	00-39	3
C400-C419	8000-8005, 8800, 8801, 8803-8805, 8810-8812, 8823, 8830, 9250, 9261-9262, 9270-9275, 9280-9282, 9290, 9300-9302, 9310-9312, 9320-9322, 9330, 9340-9342, 9363-9365, 9370-9372	00-39	3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Bone tumors, Toronto Staging is based on the presence or absence of mets and is collected in Pediatric Mets.

- Pediatric Primary Tumor and Pediatric Regional Nodes are collected for surveillance purposes

8a-8e: Malignant Bone Tumors

9623: Pediatric Primary Tumor

Coding Instructions and Codes

Note: The cortex of a bone is the dense outer shell that provides strength to the bone; the spongy center of a bone is the cancellous portion. The periosteum of the bone is the fibrous membrane covering of a bone that contains the blood vessels and nerves; the periosteum is similar to the capsule on a visceral organ.

Code	Description	SS2018 T
100	Appendicular (C400-C403, C408-C411, C413, C418-C419) • Confined to cortex of bone • Extension beyond cortex to periosteum (no break in periosteum) Spine (C412) • Confined to spine, NOS (number of segments involved not known) • Involvement of single or multiple adjacent vertebral segment(s) Pelvis (C414) • Confined to pelvis, NOS (number of segments involved not known and WITHOUT or UNKNOWN if extraosseous extension) • One to four pelvic segments involved WITHOUT or UNKNOWN if extraosseous extension (see code 200 for extraosseous extension) Localized, NOS	L
200	All sites • Extraosseous extension (beyond periosteum to surrounding tissues, including adjacent skeletal muscle(s)) Appendicular (C400-C403, C408-C411, C413, C418-C419) • Adjacent bone/cartilage Spine (C412) • Involvement of multiple non-adjacent vertebral segments • Spinal canal Pelvis (C414) • One to four pelvic segments involved WITH extraosseous extension	RE
500	Appendicular (C400-C403, C408-C411, C413, C418-C419) • Discontinuous tumors in the primary bone site ("skip" metastasis) • Skin Spine (C412) • Gross vascular invasion • Tumor thrombus in great vessels Pelvis (C414) • Encasement of external iliac vessels • Gross tumor thrombus in major pelvic vessels • Sacral neuroforamen • Sacroiliac joint	D
800	No evidence of primary tumor	U

Code	Description	SS2018 T
999	Unknown; extension not stated Primary tumor cannot be assessed Not documented in medical record Death Certificate Only	U

8a-8e: Malignant Bone Tumors

9624: Pediatric Regional Nodes

Coding Instructions and Codes

Note 1: Code only regional nodes and nodes, NOS, in this field. Distant nodes are coded in Pediatric Mets.

Note 2: Regional lymph nodes are defined as those in the vicinity of the primary tumor.

Note 3: Regional lymph node involvement is rare. If there is no mention of lymph node involvement clinically, assume that lymph nodes are negative.

Note 4: Code 800 if regional lymph nodes are involved, but there is no indication which ones are involved.

Code	Description	SS2018 N
000	No regional lymph node involvement	NONE
800	Regional lymph node(s), NOS Lymph node(s), NOS	RN
999	Unknown; regional lymph node(s) not stated Regional lymph node(s) cannot be assessed Not documented in medical record Death Certificate Only	U

8a-8e: Malignant Bone Tumors

9625: Pediatric Mets

Coding Instructions and Codes

Note: Use code 70 when the only information is "distant metastasis, NOS," and there is no documentation regarding the specific metastases.

- If there are specific metastasis documented that are not listed in codes 10, 30, or 50, assign code 50 for "other specified distant metastasis."

Code	Description	SS2018 M
00	No distant metastasis	None
10	Lung	D
30	Distant lymph node(s), NOS WITH or WITHOUT lung metastasis	D
50	Bone (other than primary site) Other specified distant metastasis WITH or WITHOUT distant lymph nodes or lung metastasis Carcinomatosis	D
70	Distant metastasis, NOS	D
99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only	None

9608: EWSR1-FLI1 fusion

Item Length: 1
NAACCR Item #: 9608
XML NAACCR ID: ewsr1Fli1Fusion
Active years: 2024+
Pediatric Schema(s):

- 6a4: Renal Tumors: Ewing Sarcoma of Kidney
- 8c: Malignant Bone Tumors: Ewing Sarcoma

Description

EWS-FLI1 fusion occurs in about 90% of Ewing Sarcomas and functions as both a pioneering transcription factor and potent oncogene.

Rationale

This is part of the National Childhood Cancer Registry (NCCR) project to collect more specific information on pediatric patients. Registries part of the NCCR will start collection on specific pediatric data items with 2024+ diagnoses.

Coding Instructions and Codes

Note 1: This SSDI is effective for diagnosis years 2024+ .

- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: EWS-FLI1 fusion occurs in about 90% of Ewing Sarcomas and functions as both a pioneering transcription factor and potent oncogene.

- Primary sources of information: molecular pathology report (may be addendum to original pathology report), CAP protocol

Note 3: Physician statement of the EWSR1-FLI1 fusion can be used to code this data item.

Code	Description
0	No gene rearrangements (fusions) identified
1	EWSR1-FLI1 gene rearrangement (fusion) present
2	EWSR1-ERG gene rearrangement (fusion) present
3	Other EWSR1 gene rearrangement (fusion) present
4	EWSR1 rearrangement present, fusion partner not known
5	Non-EWSR1 variant translocation present
7	Test ordered, results not in chart
8	Not applicable: Information not collected for this case (If this information is required by your standard setter, use of code 8 may result in an edit error.)
9	Not documented in medical record Cannot be determined by pathologist EWSR1 gene arrangements not assessed or unknown if assessed
<BLANK>	N/A - Diagnosis year is prior to 2024

9a: Rhabdomyosarcoma

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C000-C809	8900-8905, 8920, 8991	00-39	3
C000-C809	8910, 8912	All ages	3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Rhabdomyosarcoma, Toronto Staging is based on Pediatric Primary Tumor, Pediatric Regional Nodes, and Pediatric Mets.

9a: Rhabdomyosarcoma

9623: Pediatric Primary Tumor

Coding Instructions and Codes

Note: Use the following resources to determine if a tumor is localized, regional, or distant (further contiguous extension).

- EOD: Review the primary site-based schema
- Summary Stage: Review the primary site-based chapter

Code	Description	SS2018 T
100	Any size tumor • Confined to site of origin • Localized, NOS	L
200	Any size tumor • Adjacent (connective) tissue, NOS • Adjacent organ(s)/structure(s), NOS • Regional, NOS	RE
700	Any size tumor • Further contiguous extension	D
800	No evidence of primary tumor	U
999	Unknown; extension not stated Primary tumor cannot be assessed Not documented in medical record Death Certificate Only	U

9a: Rhabdomyosarcoma

9624: Pediatric Regional Nodes

Coding Instructions and Codes

Note 1: Code only regional nodes, and nodes, NOS in this field. Distant nodes are coded in Pediatric Mets.

Note 2: Use the following resources to determine if involved lymph nodes are regional or distant.

- EOD: Review the primary site-based schema
- Summary Stage: Review the primary site-based chapter

Note 3: Regional lymph node involvement is rare. For this Pediatric Schema, if there is no mention of lymph node involvement clinically, assume that lymph nodes are negative. Code 999 (unknown) only when there is no available information on the patient's disease, for example when a lab only case is abstracted from a biopsy report and no clinical information is provided.

Code	Description	SS2018 N
000	No regional lymph node involvement	NONE
800	Regional lymph node(s), NOS Lymph node(s), NOS	RN
888	Not applicable: Primary site C420-C424, C700-C729, C751-C753, C760-C809	U
999	Unknown; regional lymph node(s) not stated Regional lymph node(s) cannot be assessed Not documented in medical record Death Certificate Only	U

9a: Rhabdomyosarcoma

9625: Pediatric Mets

Coding Instructions and Codes

Note: Use the following resources to determine if involved lymph nodes are distant or if involved organs are metastatic.

- EOD: Review the primary site-based schema
- Summary Stage: Review the primary site-based chapter

Code	Description	SS2018 M
00	No distant metastasis	None
10	Distant lymph node(s), NOS	D
70	Carcinomatosis Distant metastasis WITH or WITHOUT distant lymph node(s) Distant metastasis, NOS	D
99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only	None

9a: Rhabdomyosarcoma

9609-FOXO1 Gene Rearrangements

Item Length: 1
NAACCR Item #: 9609
XML NAACCR ID: foxo1GeneRearrangements
NAACCR Alternate Name: FKHR-PAX3 or FKHR-PAX7
Active years: 2024+
Pediatric Schema(s):

- 9a: Rhabdomyosarcoma

Description

FOXO1 gene rearrangement fusions are found to be positive in about 85% of alveolar rhabdomyosarcoma patients, while are generally negative for embryonal rhabdomyosarcomas. The presence of these fusions indicates a poor prognosis. Identify these fusions will also provide new therapeutic opportunities for the treatment of fusion positive rhabdomyosarcomas (FP-RMS).

Rationale

This is part of the National Childhood Cancer Registry (NCCR) project to collect more specific information on pediatric patients, specifically for Rhabdomyosarcoma. Registries part of the NCCR will start collection on specific pediatric data items with 2024+ diagnoses.

Coding Instructions and Codes

Possible sources of information: molecular pathology reports (may be addendum to original pathology report)

Note 1: This SSDI is effective for diagnosis years 2024+.

- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: FOXO1 gene rearrangement fusions are found to be positive in about 85% of alveolar rhabdomyosarcoma patients, while are generally negative for embryonal rhabdomyosarcomas. The presence of these fusions indicates a poor prognosis. Identify these fusions will also provide new therapeutic opportunities for the treatment of fusion positive rhabdomyosarcomas (FP-RMS).

- Primary sources of information: molecular pathology report (may be addendum to original pathology report), CAP protocol

Note 3: Physician statement of FOXO1 gene rearrangements can be used to code this data item.

Note 4: This test is almost always done for Alveolar Rhabdomyosarcomas. Embryonal Rhabdomyosarcomas are usually negative, and therefore the test is usually not done.

Code	Description
0	No gene rearrangements (fusions) identified
1	FOXO1-PAX3 gene rearrangement (fusion) present
2	FOXO1-PAX7 gene rearrangement (fusion) present
3	FOXO1-PAX3 and FOXO1-PAX7 gene rearrangements (fusions) present
4	FOXO1 gene rearrangement present, fusion partner not known
7	Test ordered, results not in chart
8	Not applicable: Information not collected for this case (If this information is required by your standard setter, use of code 8 may result in an edit error.)

Code	Description
9	Not documented in medical record Cannot be determined by pathologist FOXO1 gene rearrangement not assessed or unknown if assessed
<BLANK>	N/A- Diagnosis year is prior to 2024

9b, 9d-9e: Non-Rhabdomyosarcoma

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C000-C809	8820, 8822, 8824-8827, 9150, 9160, 9491, 9540-9571, 9580	00-39	3
C000-C399, C440-C768, C809	8587, 8710-8713, 8806, 8810-8811, 8813-8815, 8821, 8823, 8830-8836, 8840-8842, 8850-8858, 8860-8862, 8870, 8880, 8881, 8890-8898, 8921, 8982, 8990, 9040-9044, 9120-9125, 9130-9133, 9135, 9136, 9141, 9142, 9161, 9170-9175, 9231, 9251, 9252, 9373, 9581	00-39	3
C000-C639, C659-C699, C739-C768, C809	8963	00-39	3
C490-C499	9180, 9210, 9220, 9240	00-39	3
C000-C399, C470-C759	9260	00-39	3
C000-C399, C470-C639, C659-C699, C739-C768, C809	9364, 9365	00-39	3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Non-Rhabdomyosarcoma, Toronto Staging is based on Pediatric Primary Tumor, Pediatric Regional Nodes, and Pediatric Mets.

9b, 9d-9e: Non-Rhabdomyosarcoma

9623: Pediatric Primary Tumor

Coding Instructions and Codes

Note: Use the following resources to determine if a tumor is localized, regional, or distant (further contiguous extension).

- EOD: Review the primary site-based schema
- Summary Stage: Review the primary site-based chapter

Code	Description	SS2018 T
100	Any size tumor • Confined to site of origin • Localized, NOS	L
200	Any size tumor • Adjacent (connective) tissue, NOS • Adjacent organ(s)/structure(s), NOS • Regional, NOS	RE
700	Any size tumor • Further contiguous extension	D
800	No evidence of primary tumor	U
999	Unknown; extension not stated Primary tumor cannot be assessed Not documented in medical record Death Certificate Only	U

9b, 9d-9e: Non-Rhabdomyosarcoma

9624: Pediatric Regional Nodes

Coding Instructions and Codes

Note 1: Code only regional nodes, and nodes, NOS in this field. Distant nodes are coded in Pediatric Mets.

Note 2: Use the following resources to determine if involved lymph nodes are regional or distant.

- EOD: Review the primary site-based schema
- Summary Stage: Review the primary site-based chapter

Note 3: Regional lymph node involvement is rare. For this Pediatric Schema, if there is no mention of lymph node involvement clinically, assume that lymph nodes are negative. Code 999 (unknown) only when there is no available information on the patient's disease, for example when a lab only case is abstracted from a biopsy report and no clinical information is provided.

Code	Description	SS2018 N
000	No regional lymph node involvement	NONE
800	Regional lymph node(s), NOS Lymph node(s), NOS	RN
888	Not applicable: Primary site C420-C424, C700-C729, C751-C753, C760-C809	NA
999	Unknown; regional lymph node(s) not stated Regional lymph node(s) cannot be assessed Not documented in medical record Death Certificate Only	U

9b, 9d-9e: Non-Rhabdomyosarcoma

9625: Pediatric Mets

Coding Instructions and Codes

Note: Use the following resources to determine if involved lymph nodes are distant or if involved organs are metastatic.

- EOD: Review the primary site-based schema
- Summary Stage: Review the primary site-based chapter

Code	Description	SS2018 M
00	No distant metastasis	None
10	Distant lymph node(s), NOS	D
70	Carcinomatosis Distant metastasis WITH or WITHOUT distant lymph node(s) Distant metastasis, NOS	D
99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only	None

10c1: Testicular

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C620-C629	9060-9065, 9070-9073, 9080-9085, 9090, 9091, 9100-9101	00-39	3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Testicular, Toronto Staging is based on Pediatric Primary Tumor, Pediatric Regional Nodes, Pediatric Mets, S Category Clinical, and S Category Pathological.

10c1: Testicular

9623: Pediatric Primary Tumor

Codes and Coding Instructions

Note 1: This schema has extension codes that are defined as "PATHOLOGICAL assessment only"

- PATHOLOGICAL assessment only codes (200, 250, 350, 450, 500) are used when there is an orchiectomy

Note 2: Code 200 for Seminomas confined to the testis.

Note 3: For codes 200 and 250, LVI [NAACCR # 1182] must be coded as none (code 0), not applicable (8), or unknown (9).

- See the STORE or SEER manual for instructions on how to code LVI

Code	Description	Pediatric T	SS2018 T
200	PATHOLOGICAL assessment only Tumour limited to the testis • Body of testis • Rete testis • Tunica albuginea • Tunica, NOS • Confined to testis, NOS Localized, NOS WITHOUT or UNKNOWN vascular/lymphatic invasion	T1	L
250	PATHOLOGICAL assessment only Epididymis • WITHOUT vascular/lymphatic invasion	T1	RE
350	PATHOLOGICAL assessment only Tumor limited to testis (including rete testis invasion) (code 200) • WITH vascular/lymphatic invasion ○ Excludes Epididymis (see code 450) Tunica vaginalis (includes implants on surface of tunica vaginalis) • WITH or WITHOUT vascular/lymphatic invasion	T2	L
450	PATHOLOGICAL assessment only Hilar soft tissue Mediastinum (of testis) Visceral mesothelial layer WITHOUT or UNKNOWN vascular/lymphatic invasion OR Epididymis WITH vascular invasion	T2	RE

Code	Description	Pediatric T	SS2018 T
500	PATHOLOGICAL assessment only Spermatic cord, ipsilateral Vas deferens	T3	RE
600	Dartos muscle, ipsilateral Scrotum, ipsilateral	T3	RE
700	Penis Scrotum, contralateral Ulceration of scrotum Further contiguous extension	T4	D
800	No evidence of primary tumor	T0	U
999	Unknown; extension not stated Primary tumor cannot be assessed Not documented in medical record Death Certificate Only	TX	U

10c1: Testicular

9624: Pediatric Regional Nodes

Codes and Coding Instructions

Note 1: Code only regional nodes and nodes, NOS, in this field. Distant nodes are coded in Pediatric Mets.

Note 2: This schema has lymph node codes that are defined as **CLINICAL** assessment only or **PATHOLOGICAL** assessment only.

- **CLINICAL** assessment only codes (100, 300) are used when there is a clinical work up only and there is no surgical resection of the primary tumor or site. This includes FNA, core biopsy, sentinel node biopsy, or lymph node excision
    - *Exception*: If patient has neoadjuvant therapy, and the clinical assessment is greater than the pathological assessment, then the clinical assessment code would take priority
- **PATHOLOGICAL** assessment only codes (200, 400, 500) are used when
    - Primary tumor or site surgically resected with
        - Any microscopic examination of regional lymph nodes. Includes
            - FNA, core biopsy, sentinel node biopsy or lymph node excision done during the clinical work up and/or
            - Lymph node dissection performed

    - Remaining codes (no designation of **CLINICAL** or **PATHOLOGICAL** only assessment) can be used based on clinical and/or pathological information

Note 3: Involvement of inguinal, pelvic, or external iliac lymph nodes WITHOUT or unknown if previous scrotal or inguinal surgery prior to presentation of the testis tumor is coded in Pediatric Mets as distant lymph node involvement.

Note 4: Regional lymph nodes include:

Aortic, NOS
- Lateral (lumbar)
- Para-aortic
- Periaortic
- Preaortic
- Retroaortic

Pericaval, NOS
- Interaortocaval
- Paracaval
- Precaval
- Retrocaval

Retroperitoneal below the diaphragm or NOS

Spermatic vein

Lymph nodes **WITH** previous scrotal or inguinal surgery
- External iliac
- Inguinal nodes, NOS
    - Deep, NOS
    - Node of Cloquet or Rosenmuller (highest deep inguinal)
    - Superficial (femoral)
- Pelvic

Note 5: Code 800 if regional lymph nodes are involved, but there is no indication which ones are involved.

Code	Description	Derived N	SS2018 N
000	No regional lymph node involvement	N0	NONE
100	CLINICAL assessment only Metastasis in lymph node(s), all less than 2 cm	N1	RN
200	PATHOLOGICAL assessment only Metastasis in lymph node(s), all less than 2 cm	Based on RNP	RN
300	CLINICAL assessment only Metastasis lymph node(s) between 2 cm and 5 cm	N2	RN
400	PATHOLOGICAL assessment only Metastasis in a lymph node, between 2 cm and 5 cm	N2	RN
500	PATHOLOGICAL assessment only Extranodal extension of lymph nodes present	N2	RN
600	Metastasis in a lymph node larger than 5 cm in greatest dimension	N3	RN
800	Regional lymph node(s), NOS Lymph node(s), NOS	N1	RN
999	Unknown; regional lymph node(s) not stated Regional lymph node(s) cannot be assessed Not documented in medical record Death Certificate Only	NX	U

10c1: Testicular

9625: Pediatric Mets

Codes and Coding Instructions

Note: Involvement of inguinal, pelvic, or external iliac lymph nodes with previous scrotal or inguinal surgery prior to presentation of the testis tumor are coded in Pediatric Regional Nodes.

Code	Description	SS2018 M
00	No distant metastasis	NONE

10	Distant lymph node(s)
	
	• WITHOUT previous scrotal or inguinal surgery OR UNKNOWN if previous scrotal or inguinal surgery
	  ○ External iliac
	  ○ Inguinal, NOS
	  ○ Deep, NOS
	  ○ Node of Cloquet or Rosenmuller (highest deep inguinal)
	  ○ Pelvis, NOS
	  ○ Superficial (femoral)
	• Retroperitoneal specified as above the diaphragm
	• Distant lymph node(s), NOS
		D

50	Lung WITH or WITHOUT distant lymph nodes	D
60	Other specified distant metastasis WITH or WITHOUT distant lymph node(s) and/or lung Carcinomatosis	D
70	Distant metastasis, NOS	D
99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only	None

10c1: Testicular

3923: S Category Clinical

Item Length: 1
NAACCR Item #: 3923
XML Parent-NAACCR ID: Tumor-sCategoryClinical
NAACCR Alternate Name: None
Active years: 2018+
Pediatric Schema(s):

- 10c1: Testicular

Description

S Category Clinical combines the results of pre-orchiectomy Alpha Fetoprotein (AFP), Human Chorionic Gonadotropin (hCG), and Lactate Dehydrogenase (LDH) into a summary S value.

Rationale

S Category Clinical is required for prognostic stage grouping in AJCC Chapter 59 *Testis*. It is a new data item for cases diagnosed 1/1/2018+. Registries part of the NCCR will start collection on specific pediatric data items with 2024+ diagnoses.

Additional Information

- For further information, refer to the **Testis** cancer protocol published by the College of American Pathologists for the AJCC Staging System *Testis*

Coding Instructions and Codes

Note 1: Code the S category as described by the physician. If the S category determined by available lab values or calculated by vendor software differs from the physician statement of the S category, the physician's statement takes precedence.

Note 2: Code the pre-orchiectomy S category according to the table below. This table is also available in AJCC 8th edition, Chapter 59, *Testis*.

- For AFP, a lab value expressed in micrograms per liter (ug/L) is equivalent to the same value expressed in nanograms per milliliter (ng/ml).

Note 3: Clinical stage values are those based on physician statement or lab values at diagnosis, prior to orchiectomy, and prior to any systemic treatment.

Note 4: All three lab values are needed for S0-S1. Only one elevated test is needed to assign S2-3. If any individual test is not available and none of the available tests results meets the S2-3 criterion for that test, assign code 9 (SX).

Code	Description
0	S0: Marker study levels within normal levels
1	S1: At least one of these values is elevated AND LDH less than 1.5 x N* AND hCG (mIU/L) less than 5,000 AND AFP (ng/mL) less than 1,000
2	S2: LDH 1.5 x N* to 10 x N* OR hCG (mIU/L) 5,000 to 50,000 OR AFP (ng/mL) 1,000 to 10,000
3	S3: Only one elevated test is needed LDH greater than 10 x N* OR hCG (mIU/mL) greater than 50,000 OR AFP (ng/mL) greater than 10,000
9	SX: Not documented in medical record S Category Clinical not assessed or unknown if assessed

*N indicates the upper limit of normal for the LDH assay.

10c1: Testicular

3924: S Category Pathological

Item Length: 1
NAACCR Item #: 3924
XML Parent-NAACCR ID: Tumor-sCategoryPathological
NAACCR Alternate Name: None
Active years: 2018+
Pediatric Schema(s):

- 10c1: Testicular

Description

S Category Pathological combines the results of post-orchiectomy Alpha Fetoprotein (AFP), Human Chorionic Gonadotropin (hCG) and Lactate Dehydrogenase (LDH) into a summary S value.

Rationale

S Category Pathological is required for prognostic stage grouping in AJCC 8th edition, Chapter 59 *Testis*. It is a new data item for cases diagnosed 1/1/2018+. Registries part of the NCCR will start collection on specific pediatric data items with 2024+ diagnoses.

Additional Information

- For further information, refer to the **Testis** cancer protocol published by the College of American Pathologists for the AJCC Staging System *Testis*

Coding Instructions and Codes

Note 1: Code the S category as described by the physician. If the S category determined by available lab values or calculated by vendor software differs from the physician statement of the S category, the physician's statement takes precedence.

Note 2: Code the post-orchiectomy S category according to the table below. This table is also available in AJCC 8th edition, Chapter 59, *Testis*.

- For AFP, a lab value expressed in micrograms per liter (ug/L) is equivalent to the same value expressed in nanograms per milliliter (ng/ml).

Note 3: Pathological stage values are those based on physician statement or lab values after orchiectomy and prior to adjuvant therapy.

Note 4: If the initial post-orchiectomy lab values remain elevated, review the subsequent tests, and use the lowest lab values (normalization or plateau) prior to adjuvant therapy or before the value rises again.

Note 5: All three lab values are needed for S0-S1. Only one elevated test is needed to assign S2-3. If any individual test is not available and none of the available tests results meets the S2-3 criterion for that test, assign code 9 (SX).

Note 6: When all the serum tumor markers are normal pre-orchiectomy and they are not repeated post-orchiectomy, code 5.

Table begins on next page

Code	Description
0	S0: Marker study levels within normal levels
1	S1: At least one of these values is elevated AND LDH less than 1.5 x N* AND hCG (mIU/L) less than 5,000 AND AFP (ng/mL) less than 1,000
2	S2 LDH 1.5 x N* to 10 x N* OR hCG (mIU/L) 5,000 to 50,000 OR AFP (ng/mL) 1,000 to 10,000
3	S3: Only one elevated test is needed LDH greater than 10 x N* OR hCG (mIU/mL) greater than 50,000 OR AFP (ng/mL) greater than 10,000
5	Post-orchiectomy serum tumor markers unknown or not done but pre-orchiectomy serum tumor markers were normal
9	SX: Not documented in medical record S Category Pathological not assessed or unknown if assessed

*N indicates the upper limit of normal for the LDH assay.

10c2: Ovarian

General Information

Primary Site	Histology	Age at Diagnosis	Behavior
C569	9060-9065, 9070-9073, 9080-9085, 9090, 9091, 9100-9101	00-39	3

Note 1: The following sources were used in the development of this pediatric schema

- Toronto Childhood Cancer Stage Guidelines, Version 2, May 2022 (IACR - Paediatric Cancer Stage Guidelines)
- SEER Extent of Disease (EOD) 2018: Codes and Coding Instructions (Extent of Disease (EOD) 2018 v3.0 (cancer.gov)
- SEER Summary Stage Manual-2018: Codes and Coding Instructions (seer.cancer.gov/tools/ssm/2018-Summary-Stage-Manual.pdf)

Note 2: For Ovarian, Toronto Staging is based on stage group only.

- Pediatric Primary Tumor, Pediatric Regional Nodes, and Pediatric Mets will be collected for surveillance purposes and will derive the Stage Group

10c2: Ovarian

9623: Pediatric Primary Tumor

Coding Instructions and Codes

Note 1: If there is involvement of the fallopian tube with no further evidence of extension, regional lymph node involvement or metastasis, and the physician verifies this is an ovary primary, code 300.

Note 2: Code 300 for extension to and/or discontinuous metastasis to any of the following pelvic organs

- Adnexa
- Adjacent (pelvic) peritoneum
- Bladder
- Bladder serosa
- Cul de sac (rectouterine pouch)
- Fallopian Tube
- Ligament(s) (broad, ovarian, round, suspensory)
- Mesosalpinx (Meosvarium)
- Parametrium
- Pelvic wall
- Rectosigmoid
- Rectum
- Sigmoid colon (including sigmoid mesentery)
- Ureter (pelvic portion)
- Uterus, NOS

Note 3: Code 400 for any evidence of peritoneal carcinomatosis, which may also be called seeding, salting, talcum powder appearance, or studding in any of the following abdominal organs (see Pediatric Mets for extraperitoneal carcinomatosis)

- Abdominal mesentery
- Diaphragm
- Gallbladder
- Intestine, large (except rectum, rectosigmoid and sigmoid colon)
- Kidneys
- Omentum (infracolic, NOS)
- Pancreas
- Pericolic gutter
- Peritoneum, NOS
- Small intestine
- Stomach
- Ureters (outside pelvis)

Code	Description	SS2018

100	Limited to one or both ovaries
	• WITH or WITHOUT tumor on ovarian surface
	• WITH or WITHOUT surgical spill
	• AND NO or UNKNOWN
	  ○ Malignant cells in ascites or peritoneal washings
	  ○ Capsule rupture
		L

Code	Description	SS2018
200	Limited to one or both ovaries • WITH Malignant Cells in ascites or peritoneal washings OR • Capsule rupture	RE
300	Tumor involves one or both ovaries with pelvic extension (below the pelvic brim) (see Note 2)	RE
400	Tumor involves one or both ovaries WITH cytologically OR histologically confirmed spread to the peritoneum outside the pelvis (see Note 3)	D
800	No evidence of primary tumor	U
999	Unknown stage: stage group not stated Stage group cannot be assessed Not documented in medical record Death Certificate Only	U

10c2: Ovarian

9624: Pediatric Regional Nodes

Coding Instructions and Codes

Note 1: Code only regional nodes and nodes, NOS, in this field. Distant nodes are coded in Pediatric Mets.

Note 2: Regional lymph nodes are defined as those in the vicinity of the primary tumor.

Code	Description	SS2018 N
000	No regional lymph node involvement	NONE

300	Intraabdominal
	Para-aortic, NOS
	• Aortic
	• Lateral aortic/lateral lumbar
	• Periaortic
	Pelvic, NOS
	• Iliac, NOS
	  ○ Common
	  ○ External
	  ○ Internal (hypogastric) (obturator)
	• Paracervical
	• Parametrial
	• Sacral, NOS
	  ○ Lateral (laterosacral)
	  ○ Middle (promontorial) (Gerota's node)
	  ○ Presacral
	  ○ Uterosacral
	Retroperitoneal, NOS
		RN

800	Regional lymph node(s), NOS Lymph node(s), NOS	RN
999	Unknown; regional lymph node(s) not stated Regional lymph node(s) cannot be assessed Not documented in medical record Death Certificate Only	U

10c2: Ovarian

9625: Pediatric Mets

Coding Instructions and Codes

Code	Description	SS2018 M
00	No distant metastasis	NONE
10	Pleural effusion with positive cytology FIGO Stage IVA	D

30	Distant lymph node(s)
	
	• Inguinal, NOS
	  ○ Inguinofemoral (groin)
	  ○ Node of Cloquet or Rosenmuller (highest deep inguinal)
	  ○ Superficial inguinal (femoral)
	
	• Distant lymph node(s)
	
	WITH or WITHOUT pleural effusion with positive cytology
		D

50	Extra-abdominal organs Liver parenchymal Spleen parenchymal Transmural involvement of intestine Carcinomatosis (involvement of multiple parenchymal organs OR diffuse involvement of multiple non-abdominal organs) • Excludes peritoneal carcinomatosis (see Pediatric Primary Tumor) WITH or WITHOUT distant lymph node(s) OR pleural effusion with positive cytology FIGO Stage IVB	D
70	Distant metastasis, NOS FIGO Stage IV [NOS]	D
99	Unknown; distant metastasis not stated Not documented in medical record Death Certificate Only	None