



Childhood Cancer Staging Rules for Population Based Registries (July 2023)



Based on the
Toronto Childhood Cancer Stage Guidelines (version 2)^{1,2}

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Abbreviations

AJCC	American Joint Committee on Cancer
APCR	Australian Paediatric Cancer Registry
COG	Children's Oncology Group
CNS	Central nervous system
CSF	Cerebrospinal fluid
FIGO	International Federation of Gynaecological Oncologists
ICCC-3	International Classification of Childhood Cancer, Third Edition
ICD-O-3	International Classification of Diseases for Oncology – Third Edition
INRGSS	International Neuroblastoma Risk Group Staging System
IRSS	International Retinoblastoma Staging System
MRI	Magnetic resonance imaging
MS	Metastatic special
NOS	Not otherwise specified
NWTSG	National Wilms tumour Study Group
PNET	Primitive neuroectodermal tumour
pPNET	Peripheral neuroectodermal tumour
RBC	Red blood cell count
RMS	Rhabdomyosarcoma
SEER	Surveillance, Epidemiology and End Results Programme
SIOP	International Society of Paediatric Oncology
TNM	Tumour, lymph nodes, metastasis
UICC	Union for International Cancer Control
WBC	White blood cell count
WHO	World Health Organisation

Introduction

The collection by population-based cancer registries of internationally consistent information on childhood cancer stage is essential for epidemiologic analysis, international benchmarking and meaningful comparisons of childhood cancer incidence and outcomes.

The TNM system is the standard staging system for most adult cancers however it is inadequate for documenting extent of disease in children. Disease-specific staging systems have been developed for childhood cancers but, for many diagnostic groups, two or more systems are in clinical use and there is no internationally uniform standard suitable for population-based cancer registration (1).

The Toronto Paediatric Cancer Stage Guidelines for population cancer registries

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. For each of a subset of major ICC-3 diagnostic groups/subgroups, the meeting reviewed all disease-specific paediatric 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 in the *Toronto Paediatric Cancer Stage Guidelines* (1).

The *Guidelines* recommended disease-specific staging systems for acute lymphoblastic leukaemia, acute myeloid leukaemia, Hodgkin lymphoma, non-Hodgkin lymphoma, neuroblastoma, renal tumours (except RCC), rhabdomyosarcoma, non-rhabdomyosarcoma soft-tissue sarcomas, osteosarcoma, Ewing sarcoma, retinoblastoma, hepatoblastoma, germ cell tumours (testicular cancer and ovarian cancer), medulloblastoma and ependymoma.

The expert group was reconvened in Lyon in 2019. At this meeting, the Toronto Guidelines were reviewed and updated by consensus (2). The main changes were that:

- a staging system was no longer considered appropriate for acute myeloid leukaemia;
- all renal tumours, with the exception of renal cell carcinomas, should use the endorsed staging systems for Wilms tumour;
- staging systems for osteosarcoma and Ewing sarcoma were combined into a single recommendation for all bone tumours;
- a staging system was endorsed for astrocytomas.

Here we provide detailed criteria for the staging systems recommended in the updated *Guidelines*. These criteria may be used by population cancer registries to derive internationally consistent and comparable information on childhood cancer stage at diagnosis using available medical records.

The *Guidelines* are endorsed by the UICC TNM Prognostic Factors project and are published in the UICC TNM Classification of Malignant Tumours 8th Edition (3).

General principles of the *Toronto Paediatric Cancer Stage Guidelines*

1. *The Guidelines are intended for use by population registries only.*

The staging systems recommended in the *Toronto Paediatric Cancer Stage Guidelines* are intended for use by population cancer registries. They are not intended to replace staging systems in clinical use nor to conflict with the stage used by clinicians in determining the treatment and prognosis of individual patients.

2. *Stage is a measure of extent of disease at diagnosis.*

The staging systems described are intended to be a measure the anatomic extent of disease at time of diagnosis. Stage is one of many prognostic indicators. Other non-anatomical prognostic indicators that are important for individual patient management and risk assessment (e.g. immuno-phenotype in leukaemia or MYCN amplification in neuroblastoma) may be collected by registries as resources permit, however, for most of the disease groups outlined here, these items do not form part of the recommended staging systems.

3. *The goal is to derive the best estimate of stage.*

The criteria provided herein are intended to enable registries to derive the best estimate of stage at diagnosis using available data sources. There are limitations inherent in collecting the data items required for staging from medical records, and assumptions may be required. However, the criteria provided here will enable a reasonable and consistent measure of stage suitable for epidemiological analysis and stratified comparisons at a population level.

4. *Resource-specific tiered staging systems are endorsed.*

The *Guidelines* endorse a two-tiered approach that provides less detailed criteria for registries with limited resources and data access (Tier 1) and more detailed staging criteria for well-resourced cancer registries (Tier 2). Tier 2 stage categories may be collapsed to Tier 1 categories to preserve comparability across registries.

This manual is concerned solely with staging at the Tier 1 level

Table 1: The Toronto Paediatric Cancer Stage Guidelines (TIER 1)

ICCC 3	Diagnostic group/subgroup	Tier 1 staging system	ICCC 3	Diagnostic group/subgroup	Tier 1 staging system
1	Acute lymphoblastic leukaemia	CNS negative	5	Retinoblastoma	Localised
		CNS positive			Regional
2b & c	Non-Hodgkin lymphoma	Limited			10c
		Advanced	Localised		
3a	Ependymoma	Localised	10c	Ovarian cancer	
		Metastatic			Metastatic
3b	Astrocytoma	Localised			10c
		Metastatic	Regional		
3c	Medulloblastoma and other CNS embryonal tumours	Localised	10c	Ovarian cancer	
		Metastatic			
6	Renal tumours (except RCCs)	Localised			10c
		Metastatic			
7	Hepatoblastoma	Localised	2a	Hodgkin lymphoma	
		Metastatic			Ann Arbor-stage IIA/B
8	Malignant bone tumours	Localised			2a
		Metastatic	Ann Arbor-stage IVA/B		
9a	Rhabdomyosarcoma	Localised	4	Neuroblastoma	
		Metastatic			Locoregional
9b,d & e	Non-rhabdomyosarcoma soft tissue sarcoma	Localised			4
		Metastatic	MS disease		

General rules of staging

1. Stage is defined as extent of disease at the time of diagnosis and is based on evidence acquired before treatment (with the exception of renal tumours, see page 18).
2. For all diagnostic groups including Wilms tumour, the presence of distant metastases is assessed clinically or pathologically before treatment.
3. If any of the information required for staging is missing from the medical record, stage is assessed as unknown. For example, if there is no record of CSF examination, leukaemia cases cannot be staged, and if there has been no bone marrow aspiration or biopsy, then lymphoma cases cannot be staged.
4. If the relevant investigations were performed and there is no mention of a data item, then it should be assumed that the item is negative/absent; for example:
 - if there is no mention of metastases then assume '*no metastases*';
 - if there is no mention of nodal involvement, then assume '*no nodal involvement*'.
5. If stage at the moment of diagnosis (before treatment by chemotherapy or radiotherapy) has already been explicitly recorded as TNM codes (or by other staging systems), the Paediatric Stage Equivalent can be determined from Table 2

Table 2: Paediatric Cancer Stage: Tier 1 and equivalents in other staging systems

Codes for 3: non-Hodgkin lymphoma			
Stage	Code	TNM	St Jude/Murphy ⁽⁵⁾
Limited	L	--	1,2,3
Advanced	A	--	4
Unknown	X		

Codes for 4: Neuroblastoma				
Stage	Code	TNM	UICC/AJCC Stage	INRGSS stage ⁽⁶⁾
Localised	L	--	--	L1
Loco-regional	LR	--	--	L2
Metastatic	M	--	--	M
MS	MS	--	--	MS
Unknown	X			

Codes for 5: Renal tumours (excluding renal cell carcinoma)					
Stage	Code	TNM	UICC/AJCC Stage	COG/NWTSG	SIOP
Localised	L	Any T, any N, M0	1,2,3	1,2,3	y1,y2,y3
Metastatic	M	Any T, any N, M1	4	4	4
Unknown	X				

Codes for 6 & 7: Soft tissue sarcomas			
Stage	Code	TNM	UICC/AJCC Stage
Localised	L	Any T, any N, M0	1,2,3
Metastatic	M	Any T, any N, M1	4
Unknown	X		

Codes for 8 Malignant bone tumours			
Stage	Code	TNM	UICC/AJCC Stage
Localised	L	Any T, N0, M0	1,2,3
		Any T, N1, M0	4b
Metastatic	M	Any T, N0, M1	4a, 4b
Unknown	X		

Codes for 9: Retinoblastoma				
Stage	Code	TNM	UICC/AJCC Stage	IRSS ⁽⁷⁾
Localised	L	T1-3, N0, M0	1	0,1,2
Regional	R	T4, N0, M0 Any T, N1, M0	2,3	3
Metastatic	M	Any T, any N, M1	4	4
Unknown	X			

Codes for 10: Liver tumours			
Stage	Code	TNM	UICC/AJCC Stage
Localised	L	T1-3, any N, M0	1,2,3
Metastatic	M	T4, any N, M0 Any T, any N, M1	4
Unknown	X		

Codes for 11: Testicular cancers			
Stage	Code	TNM	UICC/AJCC Stage
Localised	L	Any T, N0, M0	1
Regional	R	Any T, any N, M0	2
Metastatic	M	Any T, any N, M1	3
Unknown	X		

Codes for 12: Ovarian cancers				
Stage	Code	TNM	UICC/AJCC Stage	FIGO
Localised	L	T1, N0, M0	1	1
Regional	R	T2-3, N0, M0 Any T, N1, M0	2,3	2,3
Metastatic	M	Any T, any N, M1	4	4
Unknown	X			

Codes for 14 & 15: CNS tumours				
Stage	Code	TNM	UICC/AJCC Stage	M stage
Localised	L	--	--	0
Metastatic	M	--	--	1,2,3,4
Unknown	X			

1. Acute lymphoblastic leukaemia

International Classification of Childhood Cancer – Third Edition (4)

1a – Lymphoid leukaemias: 9811-9818, 9820, (9823), 9826, 9827, 9831–9837, 9940, 9948

ICD-O-3

Sites: C00.0-C80.9

Information required for staging
- Clinical signs of CNS involvement (see definitions and notes) - Blasts in the cerebrospinal fluid (CSF) from cytospin.

Definitions and notes
<p><u>CSF reports</u></p> <ul style="list-style-type: none"> - Cytospin is required to determine the presence or absence of blasts in the CSF. - If blasts are referred to as “occasional” or “seen” or similar wording, assume blasts are present. - If there is no mention of blasts, assume blasts are absent. <p><u>Clinical signs of CNS involvement are defined as</u></p> <ul style="list-style-type: none"> - Radiologic evidence of intracranial, intradural mass. - Cranial nerve palsy (e.g. facial weakness, ptosis), brain/eye involvement or hypothalamic syndrome. <p>Extraocular orbital masses, severe headaches and eye swelling (in the absence of signs of cranial nerve involvement) are not sufficient to constitute CNS involvement.</p>

Staging rules for acute lymphoblastic leukaemia	
CNS-	<ul style="list-style-type: none"> • No clinical signs of CNS involvement <p style="text-align: center;"><i>and</i></p> <ul style="list-style-type: none"> • no blasts in CSF *
CNS+	<ul style="list-style-type: none"> • Clinical signs of CNS involvement <p style="text-align: center;"><i>or</i></p> <ul style="list-style-type: none"> • blasts in CSF *

Database entry codes for acute lymphoblastic leukaemia	
Stage	Code
CNS -	C-
CNS+	C+
Unknown	X

* Staging cases with missing information

	Involvement of CNS (clinical signs)	Blasts in CSF	STAGE
1	?	?	X
2	-	?	X
3	?	-	X
4	-	-	CNS-
5	+	?	CNS+
6	?	+	CNS+
7	+	-	CNS+
8	-	+	CNS+
9	+	+	CNS+

+ Evidence of presence - Absent ? No information / not examined

2. Hodgkin lymphoma

International Classification of Childhood Cancer – Third Edition

2a - Hodgkin Lymphoma: 9650-9655, 9659, 9661-9665, 9667

ICD-O-3

Sites: C00.0-C80.9

Staging is identical to the Ann Arbor staging system (8).

Information required for staging

- Constitutional symptoms (see definitions and notes)
- Diffuse or disseminated (multifocal) involvement of one or more extra-lymphatic organs
- Distant disease: isolated (non-contiguous) extra-lymphatic organ involvement – Involvement of liver
- Involvement of lungs - Bone marrow involvement, from bone marrow aspirate or biopsy
- CSF involvement, from CSF examination
- The number of lymph node regions involved, above and below the diaphragm, from imaging. Lymph node regions are listed in Figures 1a and 1b
- The number of extra-lymphatic organs or sites involved, above and below the diaphragm, from imaging.

Definitions and notes

Constitutional symptoms

The suffix A or B is added to the stage according to the absence or presence of defined constitutional symptoms, as follows:

A = no constitutional symptoms are recorded, or the medical record states there are no constitutional symptoms

B = medical record states there are constitutional symptoms

Constitutional symptoms are:

- *Fevers.* Unexplained fever with temperature above 38 degrees Celsius (100.4 degrees F).
- *Night sweats.* Drenching sweats (e.g. those that require change of bedclothes).
- *Weight loss.* Unexplained weight loss of more than 10% of usual body weight in the 6 months prior to diagnosis.

Lymphatic regions above the diaphragm

1. Waldeyer's Ring

- Oropharynx (palatine tonsil)
- Nasopharynx (palatine tonsil)
- Adenoids
- Base of tongue (lingual tonsil)

2. Cervical, supraclavicular, occipital and pre-auricular

- Auricular lymph node
- Cervical lymph node
- Facial lymph node
- Jugular lymph node
- Mandibular lymph node
- Occipital lymph node
- Parotid lymph node
- Preauricular lymph node
- Prelaryngeal lymph node
- Pretracheal lymph node
- Retropharyngeal lymph node
- Scalene lymph node
- Sublingual lymph node
- Submandibular lymph node
- Submaxillary lymph node
- Submental lymph node
- Supraclavicular lymph node

3. Infraclavicular

- Infraclavicular lymph node
- Subclavicular lymph node

4. Axillary and pectoral lymph nodes

- Axillary lymph node
- Pectoral lymph node
- Subscapular lymph node
- Internal mammary

5. Epitrochlear and brachial

- Brachial lymph node
- Cubital lymph node
- Epitrochlear lymph node

6. Hilar

- Bronchial lymph node
- Bronchopulmonary lymph node
- Hilar lymph node
- Pulmonary hilar lymph node

7. Mediastinal

- Diaphragmatic lymph node
- Esophageal lymph node
- Innominate lymph node
- Intercostal lymph node
- Mediastinal lymph node
- Parasternal lymph node
- Paratracheal lymph node
- Peritracheal lymph node
- Pulmonary lymph node, NOS
- Thoracic lymph node
- Tracheal lymph node
- Tracheobronchial lymph node
- Subaortic lymph node (para-aortic)

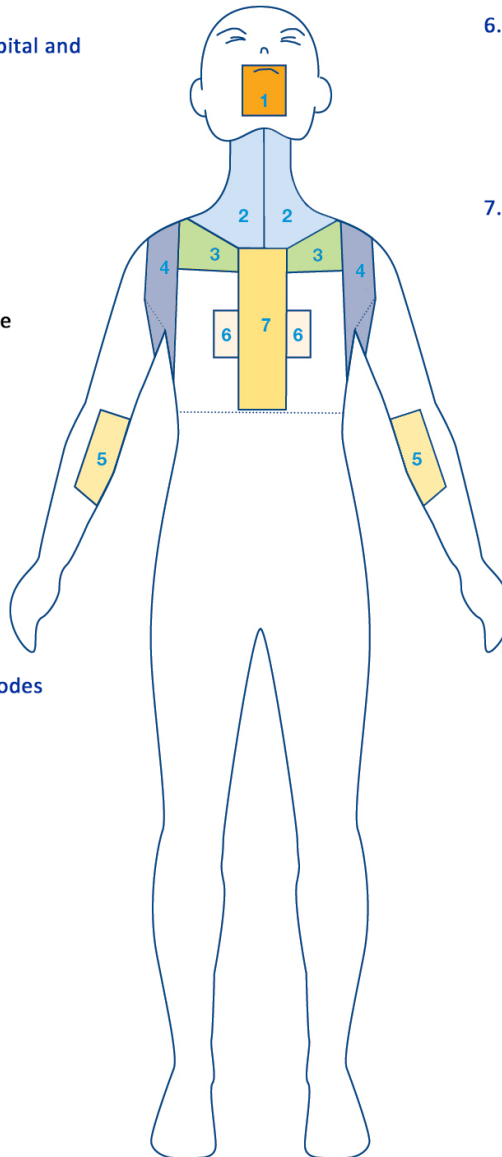


Figure 1a: Lymphatic regions above the diaphragm for the staging of Hodgkin and Non-Hodgkin Lymphoma

Lymphatic regions below the diaphragm

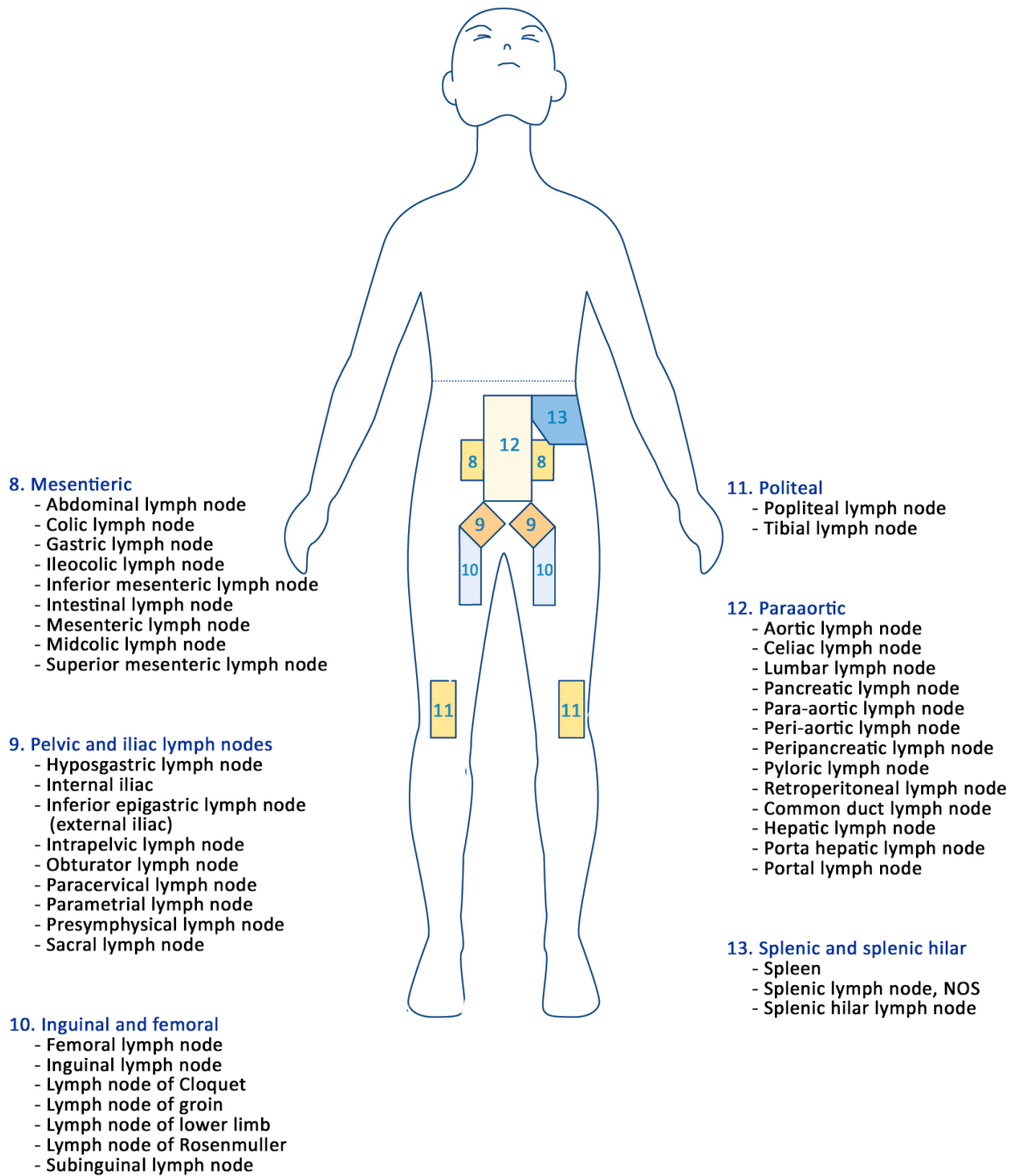


Figure 1b: Lymphatic regions below the diaphragm for the staging of Hodgkin and Non-Hodgkin Lymphoma

Staging criteria for Hodgkin lymphoma

The suffix **A** or **B** is added to the stage according to the absence or presence of defined constitutional symptoms, as follows:

A = no constitutional symptoms are recorded, or the medical record states there are no constitutional symptoms

B = medical record states there are constitutional symptoms

Constitutional symptoms are:

- *Fevers.* Unexplained fever with temperature above 38 degrees Celsius (100.4 degrees F).
- *Night sweats.* Drenching sweats (e.g. those that require change of bedclothes).
- *Weight loss.* Unexplained weight loss of more than 10% of usual body weight in the 6 months prior to diagnosis.

TIER 1

Stage I

- Involvement of a single lymph node region
- or
- Involvement of a single extralymphatic organ or site, without lymph node involvement.

Stage II

- Involvement of two or more lymph node regions on the SAME side (either above or below) of the diaphragm
- or
- Localised involvement of a single extralymphatic organ or site in association with regional lymph node involvement (i.e. local extension from a lymph node area into a nearby organ), *with or without* involvement of other contiguous lymph node regions on the SAME side (either above or below) of the diaphragm.

Stage III

- Involvement of lymph node regions on BOTH sides (above and below) of the diaphragm. This may be accompanied by:
 - extra-lymphatic extension in association with adjacent lymph node involvement (i.e. local extension from a lymph node area into a nearby organ) and/or
 - involvement of spleen.

Stage IV

- Diffuse or disseminated (multifocal) involvement of one or more extra lymphatic organs with or without associated lymph node involvement or
- Isolated (non-contiguous) extra-lymphatic organ involvement in the absence of adjacent regional lymph node involvement, but in conjunction with disease in distant site(s). or
- Any involvement of liver, bone marrow, lungs (except by direct extension from another site) or CSF.

Database entry codes for Hodgkin lymphoma	
Stage	Code
Stage IA	1A
Stage IB	1B
Stage IIA	2A
Stage IIB	2B
Stage IIIA	3A
Stage IIIB	3B
Stage IVA	4A
Stage IVB	4B
Unknown	X

3. Non-Hodgkin lymphoma

International Classification of Childhood Cancer – Third Edition

2b – Non-Hodgkin lymphomas (except Burkitt lymphoma): 9591, 9670, 9671, 9673, 9675, 9678–9680, 9684, 9688–9691, 9695, 9698–9702, 9705, 9708, 9709, 9714, 9716–9719, 9724, 9727–9729, 9731–9735, 9737-9738, 9760–9762, 9764–9769, 9823, 9827, 9970

2c – Burkitt lymphoma: 9687

ICD-O-3

Sites: C00.0-C80.9 (unless otherwise specified and excluding cutaneous lymphomas, C44_)

Information required for staging
- Central nervous system (CNS) involvement (see definitions and notes).
- Bone marrow involvement, from bone marrow aspiration or biopsy

Definitions and notes
CNS is considered involved in case of: <ul style="list-style-type: none"> Any CNS tumour mass (identified by imaging techniques [i.e., CT, MRI]), or Cranial nerve palsy that cannot be explained by extradural lesions, or Blasts morphologically identified in CSF (in the absence of a CNS tumour mass and cranial nerve palsy, a CSF report is required to confirm or exclude CNS involvement).
BM involvement: Morphologic evidence of ≥ 5% blasts or lymphoma cells by BM aspiration or biopsy.

Staging rules for non-Hodgkin lymphoma	
Limited	No involvement of CNS <u>and</u> no involvement of bone marrow*
Advanced	Involvement of CNS and / or bone marrow*

Database entry codes for non-Hodgkin lymphoma	
Stage	Code
Limited	L
Advanced	A
Unknown	X

* Staging cases with missing information (?)

Case	Bone Marrow	CNS	STAGE
1	?	?	X
2	-	?	X
3	?	-	X
4	-	-	L
5	+	?	A
6	?	+	A
7	+	-	A
8	-	+	A
9	+	+	A

+Positive/present - Negative/absent ?no information

4. Neuroblastoma

International Classification of Childhood Cancer – Third Edition

4a – Neuroblastoma and Ganglioneuroblastoma; 9490, 9500

ICD-O-3

Sites: C00.0-C80.9

Information required for staging
<ul style="list-style-type: none"> - Age of the case in months/days - Distant metastatic disease - Site of metastases (skin, liver or bone marrow) - Locoregional spread

Staging rules for neuroblastoma	
Localised	Localised tumour not involving vital structures and confined to one body compartment (neck, chest, abdomen, pelvis).
Locoregional	Tumour with local spread to more than one body compartment
Metastatic	<p>Distant metastatic disease (i.e., not contiguous with the primary tumour) except as defined for stage MS.</p> <p>Nonregional (distant) lymph node involvement is metastatic disease. However, an upper abdominal tumour with enlarged lower mediastinal nodes or a pelvic tumour with inguinal lymph node involvement is considered locoregional disease.</p> <p>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 tumour.</p>
MS	Metastatic disease confined to skin, liver, and/or bone marrow in a patient less than 18 months (547 days).

Database codes for neuroblastoma	
Stage	Code
Limited	L
Loco regional	LR
Metastatic	M
MS	MS
Unknown	X

5. Renal tumours (excluding renal cell carcinomas)

International Classification of Childhood Cancer – Third Edition

6a – Nephroblastoma; 8959, 8960, 8963, 8964-8967, 9364

6c – Unspecified malignant renal tumours; 8000-8005

ICD-O-3

Sites: C64.9

Information required for staging
- Distant metastases

Definitions and notes
<p>In cases of bilateral disease</p> <ul style="list-style-type: none"> - the presence of synchronous disease should be noted (LATERALITY = 4) - for purpose of staging, only the most advanced kidney should be recorded. <p>At diagnosis, if diagnostic imaging reports on the status of the liver, lung, bone, brain and other sites and mentions the words “suspicious”, “highly suspicious”, “possible” or “highly suspected”, record as metastatic disease (M) regardless of upfront surgery or chemotherapy.</p> <p>Note that the majority of renal tumours in childhood are Wilms tumours.</p>

Staging rules for renal tumours (excluding renal cell carcinomas)	
Localised	Tumour confined to the area of origin including abdominal lymph nodes
Metastatic	Distant metastases present at diagnosis

Database entry codes for renal tumours	
Stage	Code
Localised	L
Metastatic	M
Unknown	X

6. Rhabdomyosarcoma

International Classification of Childhood Cancer – Third Edition

9a – Rhabdomyosarcomas: 8900-8905, 8910, 8912, 8920, 8991

ICD-O-3

Sites: C00.0-C80.9

Information required for staging
- Distant metastases

Staging rules for rhabdomyosarcoma	
Localised	Tumour confined to the area of origin including the regional lymph nodes.
Metastatic	Distant metastases present

Database entry codes for rhabdomyosarcoma	
Stage	Code
Localised	L
Metastatic	M
Unknown	X

7. Non-rhabdomyosarcoma soft-tissue sarcoma

International Classification of Childhood Cancer – Third Edition

9b – Fibrosarcomas, peripheral nerve sheath tumours, and other fibrous neoplasms: 8810, 8811, 8813–8815, 8821, 8823, 8834–8835 8820, 8822, 8824–8827, 9150, 9160, 9491, 9540–9571, 9580

9d – Other specified soft tissue sarcomas: 8587, 8710–8713, 8806, 8830–8833, 8836, 8840–8842, 8850–8858, 8860–8862, 8870, 8880, 8881, 8890–8898, 8921, 8963, 8982, 8990, 9040–9044, 9120–9125, 9130–9133, 9135, 9136, 9141, 9142, 9161, 9170–9175, 9210, 9220, 9231, 9240, 9251, 9252, 9260, 9364, 9365, 9373, 9581

9e – Unspecified soft tissue sarcomas 8800–8805

* Morphology 9180/3 is not included with 9d, but is in Osteosarcoma group

ICD-O-3

Sites: C00.0-C80.9 (unless otherwise specified)

Information required for staging
- Distant metastases

Staging rules for non- rhabdomyosarcoma soft tissue sarcomas	
Localised	Tumour confined to the area of origin including the regional lymph nodes.
Metastatic	Distant metastases present

Database entry codes for non-rhabdomyosarcoma soft tissue sarcomas	
Stage	Code
Localised	L
Metastatic	M
Unknown	X

8. Malignant bone tumours

International Classification of Childhood Cancer – Third Edition

8a – Osteosarcoma: 9180-9187, 9191-9195, 9200

8b – Chondrosarcomas: 9210, 9220, 9240, 9221, 9230, 9241-9243

8c – Ewing tumour and related sarcomas of bone: 9260, 9363-9365

8d – Other specified malignant bone tumours: 8810, 8811, 8823, 8830, 8812, 9250, 9261, 9262, 9270–9275, 9280–9282, 9290, 9300–9302, 9310–9312, 9320–9322, 9330, 9340–9342, 9370–9372

8e – Unspecified malignant bone tumours: 8000–8005, 8800, 8801, 8803–8805

ICD-O-3

Sites: C40.0-C41.9, C76.0-C76.8, C80.9

Information required for staging

- Distant metastases

Definitions and notes

“Skip lesions”, “skip metastases” or “seeding” in the same bone as the primary tumour are considered Localised and not metastatic; if in a different bone to the primary tumour these are considered metastatic.

Staging rules for malignant bone tumours

Localised	Tumour confined to the area of origin including regional lymph nodes
Metastatic	Distant metastases present

Database entry codes for malignant bone tumours

Stage	Code
Localised	L
Metastatic	M
Unknown	X

9. Retinoblastoma

International Classification of Childhood Cancer – Third Edition

5 – Retinoblastoma: 9510-9514

ICD-O-3

Sites: C69.0-69.9

Information required for staging
- Distant metastases - Involvement of the orbit - Involvement of regional lymph nodes

Definitions and notes
In cases of bilateral disease: <ul style="list-style-type: none">- the presence of synchronous disease should be noted (LATERALITY = 4)- for purpose of stage, only the most advanced eye should be recorded.- Intraocular disease includes involvement of the optic nerve to the transection line

Staging rules for retinoblastoma	
Localised	Intraocular
Regional	Orbital extension or regional lymph nodes
Metastatic	Distant metastases present

Database entry codes for retinoblastoma	
Stage	Code
Localised	L
Regional	R
Metastatic	M
Unknown	X

10. Hepatoblastoma

International Classification of Childhood Cancer – Third Edition

7a – Hepatoblastoma: 8970

ICD-O-3

Site: C22.0

Information required for staging
- Distant metastases

Staging rules for hepatoblastoma	
Localised	Tumour confined to the area of origin including regional lymph nodes
Metastatic	Distant metastases present

Database entry codes for hepatoblastoma	
Stage	Code
Localised	L
Metastatic	M
Unknown	X

11. Testicular germ cell tumours

International Classification of Childhood Cancer – Third Edition

10c – Malignant gonadal germ cell tumours: 9060-9065, 9070-9073, 9080-9085, 9090-9091, 9100, 9101

ICD-O-3

Sites: C62.0-C62.9

Information required for staging
- Distant metastases - Involvement of regional lymph nodes

Staging rules for testicular cancer	
Localised	Tumour confined to the testes
Regional	Tumour extension to regional lymph nodes: <ul style="list-style-type: none"> - Interaortocaval - Para-aortic (periaortic) - Paracaval - Preaortic - Precaval - Retroaortic - Retrocaval - Along spermatic cord
Metastatic	Distant metastases present

Database entry codes for testicular cancer	
Stage	Code
Localised	L
Regional	R
Metastatic	M
Unknown	X

12. Ovarian germ cell tumours

International Classification of Childhood Cancer – Third Edition

10c – Malignant gonadal germ cell tumours: 9060-9065, 9070-9073, 9080-9085, 9090-9091, 9100, 9101

ICD-O-3

Site: C56.9

Information required for staging
- Distant metastases - Involvement of retroperitoneal lymph nodes - Extent of tumour

Staging rules for ovarian cancer	
Localised	Tumour confined to ovaries
Regional	Tumour involves one or both ovaries with pelvic extension and/or spread to the peritoneum outside the pelvis and/or retroperitoneal lymph nodes
Metastatic	Distant metastatic disease <u>excluding</u> peritoneal metastases

Database entry codes for ovarian cancer	
Stage	Code
Localised	L
Regional	R
Metastatic	M
Unknown	X

13. Astrocytomas

International Classification of Childhood Cancer – Third Edition

3b – astrocytomas: 9384, 9400–9411, 9420, 9421–9424, 9440–9442*

* Includes tumours with non-malignant behaviour for all morphology codes shown

ICD-O-3

Sites: C71-0 – 72.9

Information required for staging
- Distant metastases

Staging rules for astrocytomas	
Localised	Localised disease
Metastatic	Disease metastases present

Database entry codes for astrocytomas	
Stage	Code
Localised	L
Metastatic	M
Unknown	X

14. Medulloblastoma and other CNS embryonal tumours

International Classification of Childhood Cancer – Third Edition

3c – Intracranial and intraspinal embryonal tumours 9362*, 9470–9472, 9473*, 9474, 9480*, 9501-9504*, 9508*

* Includes tumours with non-malignant behaviour for all morphology codes shown

ICD-O-3

Sites: C70.0-C72.9, C75.3

Information required for staging
- Distant metastases - Tumour cells in the cerebrospinal fluid

Staging rules for medulloblastoma	
Localised	Localised disease
Metastatic	Disease beyond local site (e.g., other lesions in brain or spine, OR tumour cells in CSF, OR distant metastases)

Database entry codes for medulloblastomes	
Stage	Code
Localised	L
Metastatic	M
Unknown	X

15. Ependymoma

International Classification of Childhood Cancer – Third Edition

3a – Ependymomas: 9383, 9391-9394*

* Includes tumours with non-malignant behaviour for all morphology codes shown

ICD-O-3

Sites: C71.0-C72.9

Information required for staging
- Distant metastases - Tumour cells in the cerebrospinal fluid

Staging rules for ependymoma	
Localised	Localised disease
Metastatic	Disease beyond local site (e.g., other lesions in brain or spine, OR tumour cells in CSF, OR distant metastases)

Database entry codes for ependymomas	
Stage	Code
Localised	L
Metastatic	M
Unknown	X

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Appnedix I. International Classification of Childhood Cancer, 3rd edition

Site Group	ICD-O-3 Histology (Type)	ICD-O-2/3 Site	Recode
I Leukemias, myeloproliferative diseases, and myelodysplastic diseases			
(a) Lymphoid leukemias	9820, 9823, 9826, 9827, 9831-9837, 9940, 9948	C000-C809	011
(b) Acute myeloid leukemias	9840, 9861, 9866, 9867, 9870-9874, 9891, 9895-9897, 9910, 9920, 9931	C000-C809	012
(c) Chronic myeloproliferative diseases	9863, 9875, 9876, 9950, 9960-9964	C000-C809	013
(d) Myelodysplastic syndrome and other myeloproliferative diseases	9945, 9946, 9975, 9980, 9982-9987, 9989	C000-C809	014
(e) Unspecified and other specified leukemias	9800, 9801, 9805, 9860, 9930	C000-C809	015
II Lymphomas and reticuloendothelial neoplasms			
(a) Hodgkin lymphomas	9650-9655, 9659, 9661-9665, 9667	C000-C809	021
(b) Non-Hodgkin lymphomas (except Burkitt lymphoma)	9591, 9670, 9671, 9673, 9675, 9678-9680, 9684, 9689-9691, 9695, 9698-9702, 9705, 9708, 9709, 9714, 9716-9719, 9727-9729, 9731-9734, 9760-9762, 9764-9769, 9970	C000-C809	022
(c) Burkitt lymphoma	9687	C000-C809	023
(d) Miscellaneous lymphoreticular neoplasms	9740-9742, 9750, 9754-9758	C000-C809	024
(e) Unspecified lymphomas	9590, 9596	C000-C809	025
III CNS and miscellaneous intracranial and intraspinal neoplasms			
(a) Ependymomas and choroid plexus tumor	9383, 9390-9394	C000-C809	031
(b) Astrocytomas	9380	C723	032
	9384, 9400-9411, 9420, 9421-9424, 9440-9442	C000-C809	032
(c) Intracranial and intraspinal embryonal tumors	9470-9474, 9480, 9508	C000-C809	033
	9501-9504	C700-C729	033

(d) Other gliomas	9380	C700-C722, C724-C729, C751, C753	034
	9381, 9382, 9430, 9444, 9450, 9451, 9460	C000-C809	034
(e) Other specified intracranial and intraspinal neoplasms	8270-8281, 8300, 9350-9352, 9360-9362, 9412, 9413, 9492, 9493, 9505-9507, 9530-9539, 9582	C000-C809	035
(f) Unspecified intracranial and intraspinal neoplasms	8000-8005	C700-C729, C751-C753	036
IV Neuroblastoma and other peripheral nervous cell tumors			
(a) Neuroblastoma and ganglioneuroblastoma	9490, 9500	C000-C809	041
(b) Other peripheral nervous cell tumors	8680-8683, 8690-8693, 8700, 9520-9523	C000-C809	042
	9501-9504	C000-C699, C739-C768, C809	042
V Retinoblastoma	9510-9514	C000-C809	050
VI Renal tumors			
(a) Nephroblastoma and other nonepithelial renal tumors	8959, 8960, 8964-8967	C000-C809	061
	8963, 9364	C649	061
(b) Renal carcinomas	8010-8041, 8050-8075, 8082, 8120-8122, 8130-8141, 8143, 8155, 8190-8201, 8210, 8211, 8221-8231, 8240, 8241, 8244-8246, 8260-8263, 8290, 8310, 8320, 8323, 8401, 8430, 8440, 8480-8490, 8504, 8510, 8550, 8560-8576	C649	062
	8311, 8312, 8316-8319, 8361	C000-C809	062
(c) Unspecified malignant renal tumors	8000-8005	C649	063
VII Hepatic tumors			
(a) Hepatoblastoma	8970	C000-C809	071
(b) Hepatic carcinomas	8010-8041, 8050-8075, 8082, 8120-8122, 8140, 8141, 8143, 8155, 8190-8201, 8210, 8211, 8230, 8231, 8240, 8241, 8244-8246, 8260-8264, 8310, 8320, 8323, 8401, 8430, 8440, 8480-8490, 8504, 8510, 8550, 8560-8576	C220, C221	072
	8160-8180	C000-C809	072
(c) Unspecified malignant hepatic tumors	8000-8005	C220, C221	073

VIII Malignant bone tumors			
(a) Osteosarcomas	9180-9187, 9191-9195, 9200	C400-C419, C760-C768, C809	081
(b) Chondrosarcomas	9210, 9220, 9240	C400-C419, C760-C768, C809	082
	9221, 9230, 9241-9243	C000-C809	082
(c) Ewing tumor and related sarcomas of bone	9260	C400-C419, C760-C768, C809	083
	9363-9365	C400-C419	083
(d) Other specified malignant bone tumors	8810, 8811, 8823, 8830	C400-C419	084
	8812, 9250, 9261, 9262, 9270-9275, 9280-9282, 9290, 9300-9302, 9310-9312, 9320-9322, 9330, 9340-9342, 9370-9372	C000-C809	084
(e) Unspecified malignant bone tumors	8000-8005, 8800, 8801, 8803-8805	C400-C419	085
IX Soft tissue and other extraosseous sarcomas			
(a) Rhabdomyosarcomas	8900-8905, 8910, 8912, 8920, 8991	C000-C809	091
(b) Fibrosarcomas, peripheral nerve sheath tumors, and other fibrous neoplasms	8810, 8811, 8813-8815, 8821, 8823, 8834-8835	C000-C399, C440-C768, C809	092
	8820, 8822, 8824-8827, 9150, 9160, 9491, 9540-9571, 9580	C000-C809	092
(c) Kaposi sarcoma	9140	C000-C809	093
(d) Other specified soft tissue sarcomas	8587, 8710-8713, 8806, 8831-8833, 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	C000-C809	094
	8830	C000-C399, C440-C768, C809	094
	8963	C000-C639, C659-C699, C739-C768, C809	094
	9180, 9210, 9220, 9240	C490-C499	094
	9260	C000-C399, C470-C759	094
	9364	C000-C399, C470-C639, C659-C699,	094

		C739-C768, C809	
	9365	C000-C399, C470-C639, C659-C768, C809	094
(e) Unspecified soft tissue sarcomas	8800-8805	C000-C399, C440-C768, C809	095
X Germ cell tumors, trophoblastic tumors, and neoplasms of gonads			
(a) Intracranial and intraspinal germ cell tumors	9060-9065, 9070-9072, 9080-9085, 9100, 9101	C700-C729, C751-C753	101
(b) Malignant extracranial and extragonadal germ cell tumors	9060-9065, 9070-9072, 9080-9085, 9100-9105	C000-C559, C570-C619, C630-C699, C739-C750, C754-C768, C809	102
(c) Malignant gonadal germ cell tumors	9060-9065, 9070-9073, 9080-9085, 9090, 9091, 9100, 9101	C569, C620-C629	103
(d) Gonadal carcinomas	8010-8041, 8050-8075, 8082, 8120-8122, 8130-8141, 8143, 8190-8201, 8210, 8211, 8221-8241, 8244-8246, 8260-8263, 8290, 8310, 8313, 8320, 8323, 8380-8384, 8430, 8440, 8480-8490, 8504, 8510, 8550, 8560-8573, 9000, 9014, 9015	C569, C620-C629	104
	8441-8444, 8450, 8451, 8460-8473	C000-C809	104
(e) Other and unspecified malignant gonadal tumors	8590-8671	C000-C809	105
	8000-8005	C569, C620-C629	105
XI Other malignant epithelial neoplasms and malignant melanomas			
(a) Adrenocortical carcinomas	8370-8375	C000-C809	111
(b) Thyroid carcinomas	8010-8041, 8050-8075, 8082, 8120-8122, 8130-8141, 8190, 8200, 8201, 8211, 8230, 8231, 8244-8246, 8260-8263, 8290, 8310, 8320, 8323, 8430, 8440, 8480, 8481, 8510, 8560-8573	C739	112
	8330-8337, 8340-8347, 8350	C000-C809	112
(c) Nasopharyngeal carcinomas	8010-8041, 8050-8075, 8082, 8083, 8120-8122, 8130-8141, 8190, 8200, 8201, 8211, 8230, 8231, 8244-8246, 8260-8263, 8290, 8310, 8320, 8323, 8430, 8440, 8480, 8481, 8500-8576	C110-C119	113
(d) Malignant melanomas	8720-8780, 8790	C000-C809	114

(e) Skin carcinomas	8010-8041, 8050-8075, 8078, 8082, 8090-8110, 8140, 8143, 8147, 8190, 8200, 8240, 8246, 8247, 8260, 8310, 8320, 8323, 8390-8420, 8430, 8480, 8542, 8560, 8570-8573, 8940, 8941	C440-C449	115
(f) Other and unspecified carcinomas	8010-8084, 8120-8157, 8190-8264, 8290, 8310, 8313-8315, 8320-8325, 8360, 8380-8384, 8430-8440, 8452-8454, 8480-8586, 8588-8589, 8940, 8941, 8983, 9000, 9010-9016, 9020, 9030	C000-C109, C129-C218, C239-C399, C480-C488, C500-C559, C570-C619, C630-C639, C659-C729, C750-C768, C809	116
XII Other and unspecified malignant neoplasms			
(a) Other specified malignant tumors	8930-8936, 8950, 8951, 8971-8981, 9050-9055, 9110	C000-C809	121
	9363	C000-C399, C470-C759	121
(b) Other unspecified malignant tumors	8000-8005	C000-C218, C239-C399, C420-C559, C570-C619, C630-C639, C659-C699, C739-C750, C754-C809	122
Not Classified by ICCO or in situ			999

