
Welcome to this NDRS training module on tumours in Children, Teenagers and Young Adults, which has been designed to help Cancer Administration staff gain a better understanding of these tumours, the data requirements and the terminology used by the clinical teams.

# Agenda

- Children, Teenagers & Young Adults (CTYA)
  - Overview
  - Diseases of CTYA patients
- Acknowledgements

This module may be paused at any time




In this module we'll briefly look at the sites where CTYA disease may arise and provide relevant information on recording specific diseases. This module can be paused at any time.

# CTYA

**In this section we will cover:**

- CTYA Overview
- Brain & CNS
- Lymphoma
- Leukaemia
- Wilms tumour
- Neuroblastoma
- Hepatoblastoma
- Retinoblastoma
- Osteosarcoma
- Soft Tissue Sarcoma




We'll start with an overview of CTYA disease…

## CTYA - Overview

What constitutes a CTYA condition? As detailed in the COSD guidance:

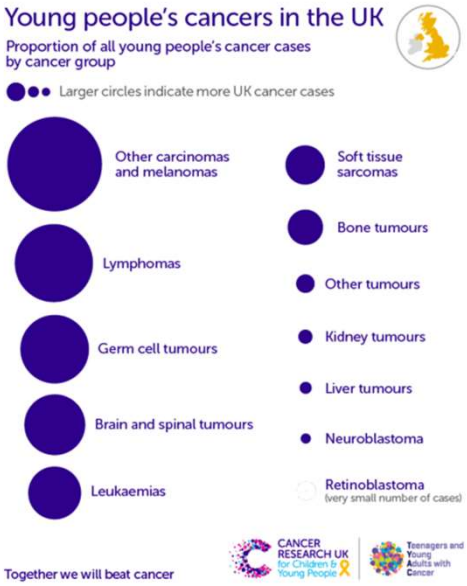
- A patient is considered as part of the CTYA cohort if they are under the age of 25 at diagnosis
  - A patient is considered to be a young adult if they are aged 19-24 inclusive at diagnosis
  - A patient is considered to be a teenager if they are aged 16-18 inclusive at diagnosis
  - A patient is considered to be a child if they are aged 0 to 15 inclusive at diagnosis (under 16)




.. The first criteria being the age ranges at which a patient is considered to be a young adult, teenager or child.

# CTYA - Overview

- CTYA may encompass many different types of cancer
- Where possible, this module will direct you to the relevant training resource for the cancer site
- Where no site-specific resource exists, basic information will be supplied, including ICD 10 coding, morphology coding and the relevant staging system (where applicable)







CTYA may encompass cancers in many different body sites. Where possible, links will be provided to the site-specific training. Where this isn't possible, basic information will be provided on recording the disease.

## CTYA - Overview

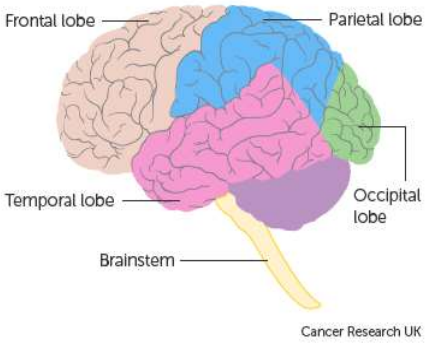
- Some cancers of children, teenagers and young adults, although still rare, are more common than others. These include:
  - Tumour of the brain / central nervous system
  - Lymphoma
  - Some types of Leukaemia
  - Wilms tumour, sometimes called a nephroblastoma (tumour of the kidney)
  - Neuroblastoma (commonly found in the abdomen, nasal cavity or adrenal glands)
  - Hepatoblastoma (tumour of the liver)
  - Retinoblastoma (tumour of the eye)
  - Osteosarcoma (tumour of the bone)
  - Soft tissue sarcoma




While cancers in children, teenagers and young adults are still rare, when they *do* arise those cancers are likely to be on this list … which we'll now look at in order

## CTYA – Brain & Central Nervous System

- For instructions on recording Brain & CNS tumours, please see the training module Brain & Central Nervous System, available here: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials
- All tumours listed within the Brain & CNS training module must be recorded in your cancer data management system, both invasive and non-invasive
- Most brain tumours are not staged, however, medulloblastomas and specified other tumours are staged using the Chang staging system. A staging data sheet for clinical use (Brain & CNS – Chang Stage) may be downloaded here: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets







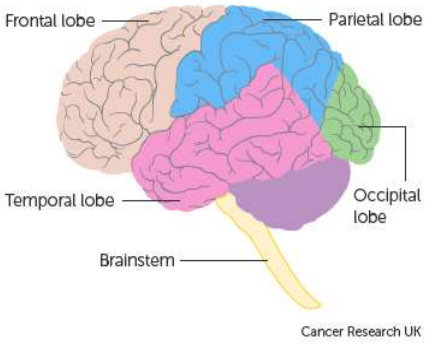
Recording of brain and central nervous system tumours is covered in a site-specific module available on the NDRS website. Please be aware that although most brain tumours are not considered stageable, medulloblastomas and other specified tumours *do* require a Chang stage. A Chang stage data sheet for clinical use may be downloaded at the link shown…

# CTYA – Brain & Central Nervous System

Chang Stage:

**CHANG STAGING SYSTEM**

| STAGE | Description |
|---|---|
| M0 | No evidence of gross subarachnoid or haematogenous metastasis. |
| M1 | Microscopic tumour cells found in cerebrospinal fluid. |
| M2 | Gross nodular seedings demonstrated in the cerebellar, cerebral subarachnoid space, or in the third or lateral ventricles |
| M3 | Gross nodular seeding in spinal subarachnoid space |
| M4 | Extraneuroaxial metastasis. |



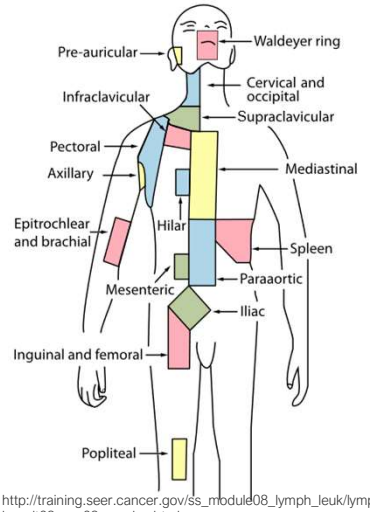




… but is included here for information

# CTYA – Lymphoma

- For specific instructions on recording Lymphomas, please see the training module Haematology Lymphoma, available here: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials
- Non-Hodgkin Lymphomas in children are staged using the Murphy St Jude classification
- Hodgkin Lymphomas in children are staged using the Ann Arbor staging system
- Morphology must be recorded for all Haematological conditions




http://training.seer.cancer.gov/ss_module08_lymph_leuk/lymph_unit02_sec02_reg_lns.html



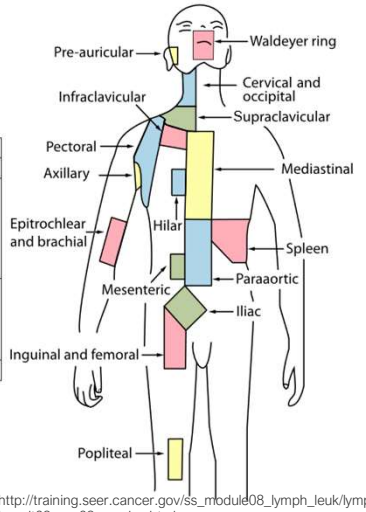

Lymphomas are covered in the Lymphoma training module. Please note that staging systems may vary depending on the type of lymphoma and the age of the patient…

# CTYA – Lymphoma

## Murphy St Jude classification

**MURPHY ST. JUDE STAGING SYSTEM**

| STAGE | Criteria for Extent of Disease |
|---|---|
| I | A single tumour (extranodal) or single anatomic area (nodal) with the exclusion of mediastinum or abdomen. |
| II | A single tumour (extranodal) with regional node involvement. Two or more nodal areas on the same side of the diaphragm. Two single (extranodal) tumours with or without regional node involvement on the same side of the diaphragm.<br>A primary gastrointestinal tract tumour, usually in the ileocecal area, with or without involvement of associated mesenteric nodes only, grossly completely resected. |
| III | Two single tumours (extranodal) on opposite sides of the diaphragm. Two or more nodal areas above and below the diaphragm. All the primary intrathoracic tumours (mediastinal, pleural, thymic). All extensive primary intraabdominal disease, unresectable. All paraspinal or epidural tumours, regardless of other tumour site(s). |
| IV | Any of the above with initial CNS and/or bone marrow involvement. |




http://training.seer.cancer.gov/ss_module08_lymph_leuk/lymph_unit02_sec02_reg_lns.html



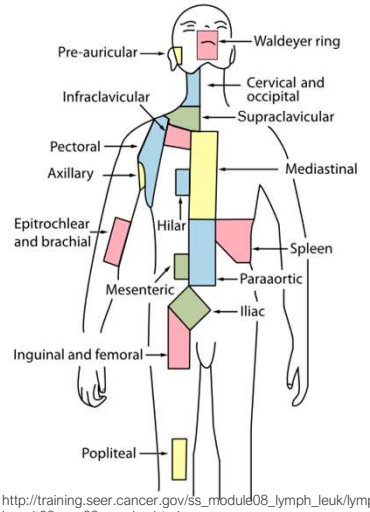

… meaning that a non-Hodgkin lymphoma in a child would be staged using the Murphy St Jude system…

# CTYA – Lymphoma

## Ann Arbor staging system

**COTSWOLDS-MODIFIED ANN ARBOR CLASSIFICATION**

| Stage | Area of involvement |
|---|---|
| Stage I | One lymph node region or extralymphatic site |
| Stage II | Two or more lymph node regions on the same side of the diaphragm |
| Stage III | Involvement of lymph node regions or structure on both side of diaphragm |
| Stage IV | Bone marrow involvement or extranodal sites beyond those designated E including multiple lung nodules and any involvement of brain or liver |
| ***Additional qualifiers*** | |
| A | No symptoms |
| B | Fever, sweats, weight loss (more than 10% body weight) |
| E | Involvement of a single extranodal site, contiguous in proximity to a known nodal site |
| X* | Bulky disease<br>Mass > 1/3 transthoracic diameter at T5 on CXR or any mass > 10cm maximum dimension |
| S | Spleen involvement |



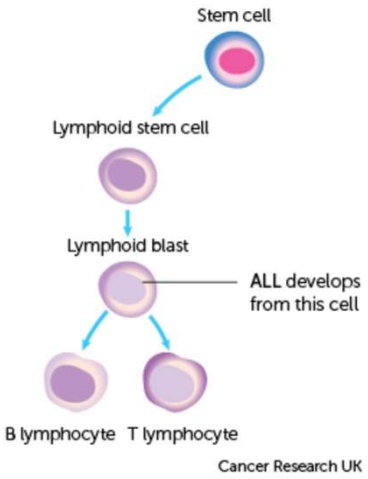

http://training.seer.cancer.gov/ss_module08_lymph_leuk/lymph_unit02_sec02_reg_lns.html




… while a Hodgkin lymphoma would be staged using the Ann Arbor staging system

## CTYA - Leukaemia

- Leukaemias developed in childhood may include Childhood Acute Lymphocytic Leukaemia (Childhood ALL) and Juvenile myelomonocytic leukaemia (JMML)
- For specific instructions on recording Leukaemia, please see the training module Haematology Leukaemia, available here: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials
- Most leukaemias are not considered stageable with the exception of chronic lymphocytic leukaemia (CLL)





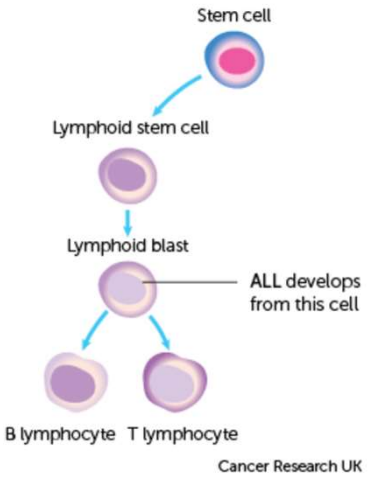


Leukaemias also have their own training module. Most leukaemias are not considered stageable with the exception of chronic lymphocytic leukaemia.

## CTYA - Leukaemia

- Binet Stage for CLL

| | |
|---|---|
| Stage A | If Platelet count > 99 x $10^9$/L and Hb >99 and 0, 1 or 2 areas of organ enlargement (number of lymph node groups plus score 1 for hepatomegaly, 1 for splenomegaly) |
| Stage B | If Platelet count> 99 and Hb>99 and 3, 4 or 5 areas of organ enlargement |
| Stage C | If Hb<100 or platelet count <100 |



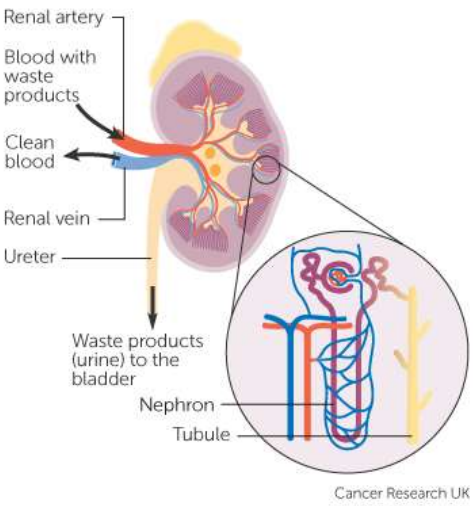




While CLL in children is *extremely* rare, the Binet stage for CLL is shown here for information

## CTYA – Wilms tumour

- For instructions on recording Wilms tumours, please see the training module Urology - Kidney, available here: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials
- Wilms tumours are staged using the Wilms staging system. A staging data sheet for clinical use may be downloaded here: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets







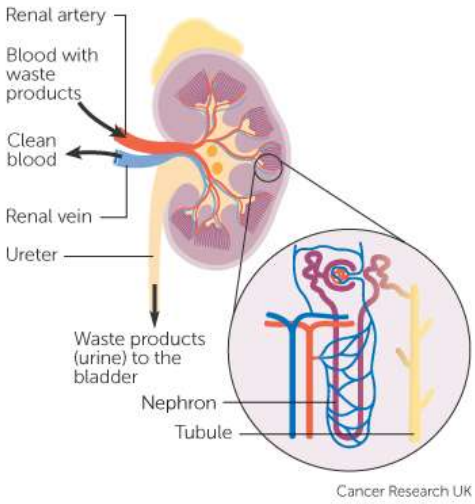
Wilms tumours are covered in the Urology – Kidney training module. A Wilms staging data sheet is available at the link shown …

# CTYA – Wilms tumour

- Wilms stage

**WILMS' TUMOUR STAGE**

| Stage | Description |
|---|---|
| Stage I | a) The tumour is limited to kidney or surrounded with a fibrous pseudocapsule if outside of the normal contours of the kidney. The renal capsule or pseudocapsule may be infiltrated with the tumour but it does not reach the outer surface, and it is completely resected (resection margins 'clear')<br>b) The tumour may be protruding ('bulging') into the pelvic system and 'dipping' into the ureter (but it is not infiltrating their walls)<br>c) The vessels of the renal sinus are not involved<br>d) Intra renal vessel involvement may be present<br>Fine needle aspiration or percutaneous core needle biopsy ('tru-cut') does not upstage the tumour.<br>The presence of necrotic tumour or chemotherapy-induced change in the renal sinus and/or within the perirenal fat should not be regarded as a reason for upstaging a tumour providing it is completely excised and does not reach the resection margins. |
| Stage II | a) The tumour extends beyond kidney or penetrates through the renal capsule and/or fibrous pseudocapsule into peri-renal fat but is completely resected (resection margins 'clear')<br>b) Tumour infiltrates the renal sinus and/or invades blood and lymphatic vessels outside the renal parenchyma but it is completely resected<br>c) Tumour infiltrates adjacent organs or vena cava but is completely resected |
| Stage III | a) Incomplete excision of the tumour which extends beyond resection margins (gross or microscopic tumour remains post-operatively)<br>b) Any abdominal lymph nodes are involved<br>c) Tumour rupture before or intra-operatively (irrespective of other criteria for staging)<br>d) The tumour has penetrated through the peritoneal surface<br>e) Tumour implants are found on the peritoneal surface<br>f) The tumour thrombi present at resection margins of vessels or ureter, transected or removed piecemeal by surgeon<br>g) The tumour has been surgically biopsied (wedge biopsy) prior to pre-operative chemotherapy or surgery.<br>The presence of necrotic tumour or chemotherapy-induced changes in a lymph node or at the resection margins is regarded as proof of previous tumour with microscopic residue and therefore the tumour is assigned stage III (because of the possibility that some viable tumour is left behind in the adjacent lymph node or beyond resection margins.) |
| Stage IV | Haematogeneous metastases (lung, liver, bone, brain, etc.) or lymph node metastases outside the abdomino-pelvic region. |
| Stage V | Bilateral renal tumours at diagnosis. Each side should be sub staged according to above classifications. |







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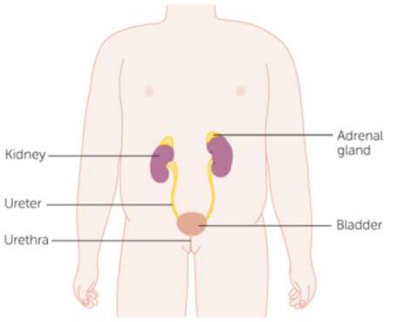
## CTYA - Neuroblastoma

- Neuroblastomas mostly affect children under the age of 5 but can sometimes develop in older children, teenagers and adults
- A neuroblastoma may develop anywhere in the body but commonly occurs in the adrenal glands or at the back of the abdomen. Neuroblastoma may also occur in the nasal cavity
- Morphology for neuroblastomas:
  - Neuroblastoma, NOS – M9500/3
  - Neuroblastoma, olfactory – M9522/3

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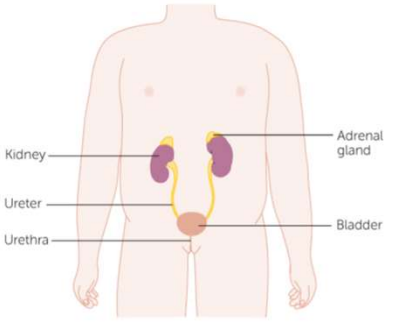
Neuroblastomas may develop anywhere in the body but tend to present in the adrenal glands, in the tissue at the back of the abdomen or in the nasal cavity. Morphology codes are shown here…

## CTYA - Neuroblastoma

- A neuroblastoma would be recorded in ICD10 using the relevant code for that body area. This includes:
  - Malignant neoplasm of adrenal gland, medulla – C74.1
  - Malignant neoplasm of other and ill-defined sites, abdomen – C76.2
  - Malignant neoplasm of nasal cavity – C30.0
- Neuroblastomas are staged using the International Neuroblastoma Risk Group Staging System (INRGSS). A staging data sheet for clinical use may be downloaded here: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets



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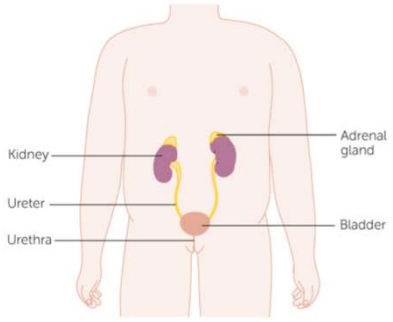
… and the relevant topographical ICD 10 codes are shown here. All neuroblastomas must be recorded in your cancer management system and require an International Neuroblastoma Risk Group stage. A Neuroblastoma staging sheet for clinical use may be downloaded from the link shown…

# CTYA - Neuroblastoma

## The International Neuroblastoma Risk Group Staging System (INRGSS):

**INTERNATIONAL NEUROBLASTOMA RISK GROUP**

| STAGE | Description |
|---|---|
| L1 | Localised tumour not involving vital structures as defined by the list of image-defined risk factors and confined to one body compartment |
| L2 | Locoregional tumour with presence of one or more image-defined risk factors |
| M | Distant metastatic disease (except stage MS) |
| MS | Metastatic disease in children younger than 18 months with metastases confined to skin, liver, and/or bone marrow |
| | |
| **Image-Defined Risk Factors (IDRFs) in Neuroblastic Tumours** | |
| Ipsilateral tumour extension within two body compartments | Neck-chest, chest-abdomen, abdomen-pelvis |
| Neck | Tumour encasing carotid and/or vertebral artery and/or internal jugular vein |
| | Tumour extending to base of skull |
| | Tumour compressing the trachea |
| Cervico-thoracic junction | Tumour encasing brachial plexus roots |
| | Tumour encasing subclavian vessels and/or vertebral and/or carotid artery |
| | Tumour compressing the trachea |
| Thorax | Tumour encasing the aorta and/or major branches |
| | Tumour compressing the trachea and/or principal bronchi |
| | Lower mediastinal tumour, infiltrating the costo-vertebral junction between T9 and T12 |
| Thoraco-abdominal | Tumour encasing the aorta and/or vena cava |
| Abdomen/pelvis | Tumour infiltrating the porta hepatis and/or the hepatoduodenal ligament |
| | Tumour encasing branches of the superior mesenteric artery at the mesenteric root |
| | Tumour encasing the origin of the coeliac axis, and/or of the superior mesenteric artery |
| | Tumour invading one or both renal pedicles |
| | Tumour encasing the aorta and/or vena cava |
| | Tumour encasing the iliac vessels |
| | Pelvic tumour crossing the sciatic notch |

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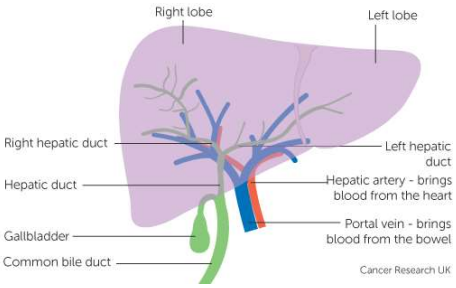
| | |
|---|---|
| Intraspinal tumour extension whatever the location provided that: | More than one third of the spinal canal in the axial plane is invaded and/or the perimedullary leptomeningeal spaces are not visible and/or the spinal cord signal is abnormal |
| Infiltration of adjacent organs/structures | Pericardium, diaphragm, kidney, liver, duodeno-pancreatic block, and mesentery |
| Conditions to be recorded, but not considered IDRFs | Multifocal primary tumours |
| | Pleural effusion, with or without malignant cells |
| | Ascites, with or without malignant cells |




… but is included here for information

## CTYA - Hepatoblastoma

- For instructions on recording hepatoblastoma, please see the training module Lower / Upper GI - Hepatobiliary, available here: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials
- Hepatoblastomas in children, teenagers and young adults are staged using the Pretext Staging System. A staging data sheet for clinical use may be downloaded here: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets





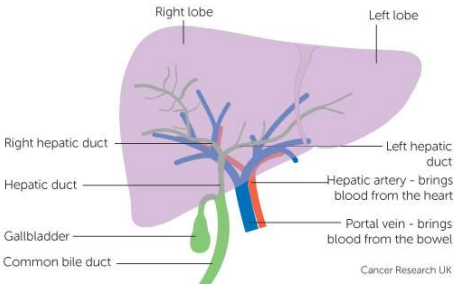


Recording of hepatoblastomas is covered in the HPB training module. Hepatoblastomas in children, teenagers and young adults require a PRETEXT stage.

# CTYA - Hepatoblastoma

**PRETEXT STAGING SYSTEM**

| PRETEXT | Definition |
|---|---|
| I | One section involved<br>Three adjoining sections are tumour free |
| II | One or two sections involved<br>Two adjoining sections are tumour free |
| III | Two or three sections involved<br>One adjoining section is tumour free |
| IV | Four sections involved |
| | |
| Annotation: | |
| V | Venous involvement, V, denotes vascular involvement of the retrohepatic vena cava or involvement of all three major hepatic veins (right, middle, and left) |
| P | Portal involvement, P, denotes vascular involvement of the main portal vein and/or both right and left portal veins |
| E | Extrahepatic involvement of a contiguous structure such as the diaphragm, abdominal wall, stomach, colon, and so on. |
| M | Distant metastatic disease (usually lungs, very rarely bone, or brain) |
| C | Caudate lobe |
| F | Multifocal tumour nodules |
| R | Tumour rupture prior to diagnosis |



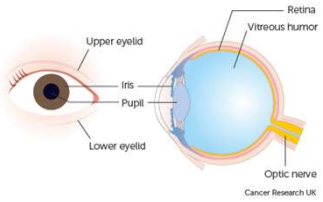




Where applicable, any annotation must be recorded for Pretext staging *in addition* to the stage

# CTYA - Retinoblastoma

Retinoblastomas develop in the retina inside the eye. They arise in immature cells that should have gone on to form part of the retina (retinoblasts). For more details on recording tumours of the Eye, please see the relevant training module: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials




Retinoblastoma are ICD10 coded to:

- Malignant neoplasm of eye and adnexa, retina – C69.2

SNOMED CT code with the ICD-O-3 morphology code in brackets:

- Retinoblastoma, NOS – M9510/3
- Retinoblastoma, differentiated – M9511/3
- Retinoblastoma, undifferentiated – M9512/3
- Retinoblastoma, diffuse – M9513/3
- Retinoblastoma, spontaneously regressed – M9514/3

The exact morphology coding may be detailed on the pathology report




Retinoblastomas develop inside the eye. Morphological type may vary but they are always ICD10 coded to C69.2

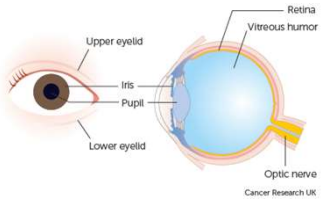
## CTYA - Retinoblastoma

Retinoblastomas are staged using the TNM staging system. A Retinoblastoma TNM staging data sheet for clinical use may be downloaded here:

https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets

Retinoblastomas must also be assessed using the appropriate assessment systems:

- Intraocular Classification of Retinoblastoma Assessment (ICRB)
- Post-treatment / extraocular assessment: International Staging System for Retinoblastoma Assessment (ISSRA)







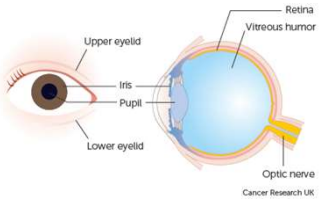
All retinoblastomas must be staged using UICC TNM v8. A Retinoblastoma staging sheet can be downloaded at the link shown.

# CTYA - Retinoblastoma

## Intraocular Classification of Retinoblastoma assessment (ICRB)

**ICRB STAGING SYSTEM**

| STAGE | Description |
|---|---|
| Group A (very low risk) | Retinoblastoma ≤3 mm (in basal dimension or thickness) |
| Group B (low risk) | Retinoblastoma >3mm (in basal dimension or thickness) or<br>• Macular location (≤3 mm to foveola)<br>• Juxtapapillary location (≤1.5 mm to disc)<br>• Additional subretinal fluid (≤3 mm from margin) |
| Group C (moderate risk) | Retinoblastoma with:<br>• Subretinal seeds ≤3 mm from tumour<br>• Vitreous seeds ≤3 mm from tumour<br>• Both subretinal and vitreous seeds ≤3 mm from tumour |
| Group D (high risk) | Retinoblastoma with:<br>• Subretinal seeds >3 mm from tumour<br>• Vitreous seeds >3 mm from tumour<br>• Both subretinal and vitreous seeds >3 mm from tumour |
| Group E (very high risk) | Extensive retinoblastoma occupying >50% globe or with:<br>• Neovascular glaucoma<br>• Opaque media from haemorrhage in anterior chamber, vitreous or subretinal space<br>• Invasion of postlaminar optic nerve, choroid (>2 mm), sclera, orbit, anterior chamber |







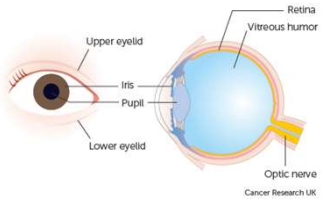
Retinoblastomas are *also* assessed for risk grouping using the ICRB system.

# CTYA - Retinoblastoma

## Post-treatment / extraocular assessment: International Staging System for Retinoblastoma Assessment (ISSRA)

**ISSRA STAGING SYSTEM**

| STAGE | Description |
|---|---|
| 0 | Patient treated conservatively |
| I | Eye enucleated, completely resected histologically |
| II | Eye enucleated, microscopic residual tumour |
| III | Regional extension |
| IIIa | Overt orbital disease |
| IIIb | Preauricular or cervical lymph node extension |
| IV | Metastatic disease |
| IVa | Haematogenous metastasis (without central nervous system involvement) |
| IVa1 | Single lesion |
| IVa2 | Multiple lesions |
| IVb | Central nervous system extension (with or without any other site of regional or metastatic disease) |
| IVb1 | Pre-chiasmatic lesion |
| IVb2 | Central nervous system mass |
| IVb3 | Leptomeningeal and cerebrospinal fluid disease |



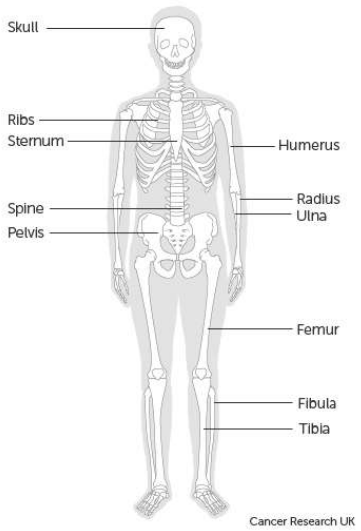




After treatment, Retinoblastomas will need an ISSRA classification.

## CTYA - Osteosarcoma

- For instructions on recording Osteosarcomas, please see the training module Sarcoma, available here: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials
- Osteosarcomas are staged using the TNM staging system. A Bone TNM staging data sheet (for clinical use) may be downloaded here: https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets







The recording of Osteosarcomas is covered in the Sarcoma training module. A staging data sheet for Bone tumours may be downloaded at the link shown.

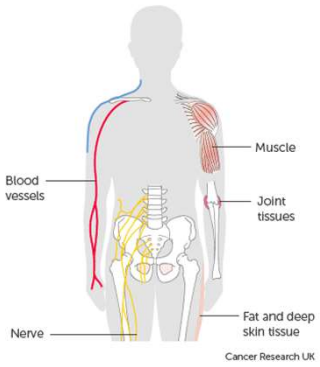
## CTYA – Soft Tissue Sarcoma

For instructions on recording Soft Tissue Sarcomas, please see the training module Sarcoma, available here:

https://digital.nhs.uk/ndrs/data/cancer-data-training-materials

Stageable soft tissue sarcomas are staged using the TNM staging system. A Rhabdomyosarcoma and Other Soft Tissue Sarcomas TNM staging data sheet (for clinical use) may be downloaded here:

https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets





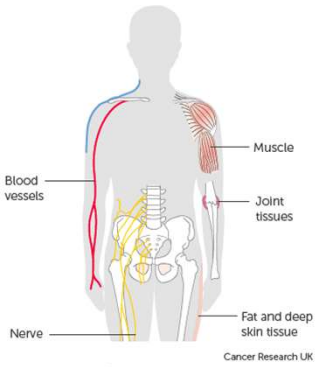


Details on recording soft tissue sarcomas can also be found in the Sarcoma training module. While not all soft tissue sarcomas are stageable, those that are will require a TNM stage. A Rhabdomyosarcoma and other soft tissue sarcomas staging sheet is available at the link shown.

# CTYA – Soft Tissue Sarcoma

In addition to a TNM stage, rhabdomyosarcomas will require an Intergroup Rhabdomyosarcoma Studies group classification:

| IRS Group | Classification |
|---|---|
| I | - Localised disease, completely resected (regional nodes not involved)<br>- Confined to muscle or organ of origin<br>- Contiguous involvement with infiltration outside the muscle or organ of origin, as through fascial planes |
| II | - Grossly resected tumour with microscopic residual disease<br>- No evidence of gross residual tumour; no evidence of regional node involvement<br>- Regional disease, completely resected (regional nodes involved and/or extension of tumour into an adjacent organ); all tumour completely resected with no microscopic residual tumour<br>- Regional disease with involved nodes, grossly resected, but with evidence of microscopic residual |
| III | - Incomplete resection or biopsy with gross residual disease |
| IV | - Distant metastatic disease present at onset (lung, liver, bones, bone marrow, brain, and distant muscle and nodes) |




A determination of favourable or unfavourable site is also required:

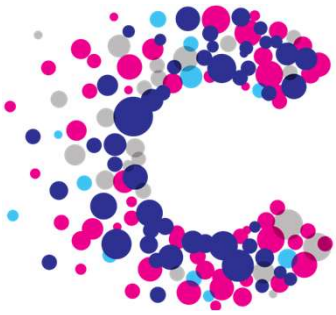
| |
|---|
| **Favourable anatomic sites**: Orbit, head and neck(excluding parameningeal tumours) and genitourinary sites (excluding bladder and prostate tumours) |
| **Unfavourable anatomic sites**: Bladder, prostate, extremity, cranial, parameningeal, trunk, retroperitoneum and all other sites not noted as favourable |




Rhabdomyosarcoma also require an IRS Group classification as well as a determination of favourable or unfavourable disease site.

## Acknowledgements

Many thanks to Cancer Research UK for the use of their images within this training module.



CANCER RESEARCH UK




We'd like to thank Cancer Research UK for the use of their images within this training module.

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If you have any questions on the information contained within this module or about COSD in general, do please feel free to email your regional Data Liaison Manager