Paediatric Rhabdomyosarcoma Histopathology Reporting Guide

**Family/Last name**

**Given name(s)**

**Patient identifiers**

**Date of birth**

**Date of request**

**Accession/Laboratory number**

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**OPERATIVE PROCEDURE**

- Not specified
- Biopsy
- Intralesional excision
- Marginal resection
- Wide local resection
- Radical resection
- Amputation, specify
- Other, specify

**TUMOUR SITE**

- Not specified
- Unknown primary site
- Bile duct
- Bladder/prostate
- Parameningeal
- Extremity
- Orbit
- Head and neck (excluding parameningeal), specify
- Genitourinary (excluding bladder/prostate), specify
- Other, specify

**TUMOUR LATERALITY**

- Left
- Right
- Not specified

**TUMOUR DIMENSIONS**

(Applicable for excision biopsies and resection specimens)

- Greatest dimension

- Additional dimensions

- Cannot be assessed, specify

**BLOCK IDENTIFICATION KEY**

(List overleaf or separately with an indication of the nature and origin of all tissue blocks)
Paediatric Rhabdomyosarcoma

HISTOLOGICAL TUMOUR TYPE (select all that apply)
(Valie list based on the World Health Organization Classification of Paediatric Tumours (2023))

- Embryonal rhabdomyosarcoma
- Alveolar rhabdomyosarcoma
- Pleomorphic rhabdomyosarcoma
- Spindle cell/sclerosing rhabdomyosarcoma
- Other, specify

LYMPH NODE STATUS

- Cannot be assessed
- No nodes submitted or found
- Number of lymph nodes examined
- Not involved
- Involved

- Number of involved lymph nodes
- Location of involved lymph nodes, specify

ANAPLASIA
(Applicable for biopsy or post-treatment resection specimens)

- Cannot be determined
- Not identified
- Present
  - Focal
  - Diffuse
  OR
  - % of cells

OTHER HISTOLOGICAL FEATURES

- None identified
- Present, specify

TREATMENT EFFECT

- No previous treatment
- No response
- Response
  - Microscopic viable tumour
  - %
  - Rhabdomyoblastic differentiation
    - Not identified
    - Present
    - % differentiation

MARGIN STATUS
(Applicable for resection specimens only)

- Cannot be assessed
- Not involved
  - Distance of tumour from closest margin
  - Specify closest margin(s) (<10 mm), if possible
- Involved
  - Specify margin(s), if possible

ANCILLARY STUDIES

- Not performed
- Performed

Immunohistochemistry

- Not performed
- MyoD1
  - Negative
  - Positive
  - %
- Myogenin
  - Negative
  - Positive
  - %
- Desmin
  - Negative
  - Positive
- Other, specify test(s) and result(s)

Gene fusion studies

- Not performed
- Pending
- No FOXO1 rearrangement
- FOXO1 rearrangement present, fusion partner not known
- PAX3::FOXO1 gene fusion
- PAX7::FOXO1 gene fusion
- Other rearrangement/fusion, specify
Paediatric Rhabdomyosarcoma

**ANCILLARY STUDIES continued**

Molecular genetic studies
- Not performed
- Pending
- MYOD1 L122R mutation
- VGLL2/NCOA2 gene fusions, specify
- EWSR1/FUS-TFCP2 gene fusion, specify
- TP53
- DICER1 mutation
- Other, specify test(s) and result(s)

Representative blocks for ancillary studies, specify those blocks best representing tumour and/or normal tissue for further study

**HISTOLOGICALLY CONFIRMED DISTANT METASTASES**

- Not applicable
- Not identified
- Present, specify site(s)

**PATHOLOGICAL STAGING**

(Applicable for resection specimens only)

**Pathologic staging system used**
- Children’s Oncology Group (COG)
- TNM

**Children's Oncology Group (COG) staging**
- Stage I Requires all of the following to be true:
  - Tumour involves favourable site (i.e., orbit, head and neck [excluding parameningeal] or genitourinary site [excluding bladder/prostate]), and; Tumour metastatic to distant site not identified
  - Tumour involves unfavourable site (i.e., bladder/prostate, extremity, parameningeal or other site not mentioned in stage I), and; Tumour size ≤5 cm, and; Tumour involvement of lymph nodes not identified, and; Tumour metastatic to distant site not identified
- Stage II Requires all of the following to be true:
  - Tumour involves unfavourable site, is ≤5 cm, and involves regional lymph nodes, but distant metastases are not identified, or; Tumour involves unfavourable site and is >5 cm, with or without regional lymph node involvement, but distant metastases are not identified

**TNM staging (UICC TNM 8th edition 2016)**

**TNM Descriptor** (only if applicable)
- y - post-therapy

**Primary tumour (pT)**
- pT0 No evidence of tumour found on histological examination of specimen
- pT1 Tumour limited to organ or tissue of origin
  - Excision complete and margins histologically free
  - pT2 Tumour with invasion beyond the organ or tissue of origin
    - Excision complete and margins histologically free
    - pT3 Tumour with or without invasion beyond the organ or tissue of origin
      - Excision incomplete
      - pT3a Evidence of microscopic residual tumour
      - pT3b Evidence of macroscopic residual tumour
      - pT3c Adjacent malignant effusion regardless of size
      - pTX\(^a\) Tumour status may not be assessed

**Regional lymph nodes (pN)**
- pN0 No evidence of tumour found on histological examination of regional lymph nodes
- pN1 Evidence of invasion of regional lymph nodes
- pN1a Evidence of invasion of regional lymph nodes
  - Involved nodes considered to be completely resected
- pN1b Evidence of invasion of regional lymph nodes
  - Involved nodes considered not to be completely resected
- pNX\(^{a,b}\) N status may not be assessed due to lack of pathological examination or inadequate information on pathological findings

\(^a\) Core for primary resection specimens (if using COG staging it is core if clinical information is available); non-core for post-treatment resection specimens.

\(^b\) Either COG or TNM can be used depending on local preference.

\(^c\) Reprinted from Pediatr Blood Cancer., Volume 69(6), Crane JN, Xue W, Qumseya A, et al. Clinical group and modified TNM stage for rhabdomyosarcoma: A review from the Children’s Oncology Group, 2022, with permission from Wiley.


\(^e\) pTX and pNX should be used only if absolutely necessary.

\(^f\) For evaluations pNX will be regarded as N0.