Phaeochromocytoma and Paraganglioma Histopathology Reporting Guide

Family/Last name

Given name(s)

Date of birth DD – MM – YYYY

Patient identifiers

Date of request DD – MM – YYYY

Accession/Laboratory number

Elements in **black text** are CORE. Elements in **grey text** are NON-CORE.

☐ indicates multi-select values ◯ indicates single select values

SCOPE OF THIS DATASET

CLINICAL INFORMATION

◯ Information not provided
◯ Information provided

Hormonal status

◯ Cannot be determined (testing status not known)
◯ Biochemically functioning (select all that apply)
- ☐ Metanephrine and/or adrenaline
- ☐ Normetanephrine and/or noradrenaline
- ☐ Methoxytyramine and/or dopamine
- ☐ Other, *specify*

◯ Biochemically silent
◯ Biochemical analysis not performed

Relevant biopsy/cytology results

◯ Information not provided
◯ No
◯ Yes, *specify*

Imaging findings

◯ Information not provided
◯ No
◯ Yes, *specify*

Previous therapy

◯ Information not provided
◯ No
◯ Yes, *specify*

Relevant familial history

◯ Information not provided
◯ No
◯ Yes, *specify*

Presence of endocrine or other tumours

◯ Information not provided
◯ No
◯ Yes, *specify*

Presence of germline mutation or familial syndrome

◯ Information not provided
◯ No
◯ Yes, *specify mutation if known*

Other clinical information, *specify*

OPERATIVE PROCEDURE (select all that apply)

◯ Not specified
☐ Biopsy (core needle, incisional, excisional), *specify*
☐ Open resection (e.g., adrenal resection, liver biopsy), *specify procedure including other organs if present*
☐ Laparoscopic
☐ Organ-sparing
☐ Other (e.g., conversion, laparoscopic to open), *specify*

SPECIMEN(S) SUBMITTED (select all that apply)

◯ Not specified
☐ Adrenalectomy
- ☐ Left ☐ Right

☐ Other resection, *specify site(s) and laterality*
☐ Biopsy tissue, *specify site(s) and laterality*

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TUMOUR FOCALITY

- ○ Unifocal
- ○ Multiple
  - Multifocal (separate tumours in the same organ), *specify number of tumours*
  - Multiple tumours in separate organs, *specify number of tumours*[a]
- ○ Cannot be assessed, *specify*

TUMOUR SITE[a] (select all that apply)

(Specify number of tumours at any site containing more than one tumour)

- ○ Not specified
- ☐ Adrenal
  - ☐ Left
  - ☐ Right
- ☐ Other abdominal or pelvic
  - ○ Paraaortic
  - ○ Urinary bladder
  - ○ Other, *specify*
- ☐ Thorax
  - ○ Paraaortic
  - ○ Cardiac
  - ○ Other, *specify*
- ☐ Head and neck
  - ○ Carotid body
    - ○ Left ○ Right
  - ○ Middle ear (jugulotympanic)
    - ○ Left ○ Right
  - ○ Vagal
    - ○ Left ○ Right
  - ○ Laryngeal
    - ○ Left ○ Right
  - ○ Other, *specify site(s) and laterality*

[a] *If multiple tumours from different organs are present, separate datasets should be used to record all following elements for each tumour.*

SPECIMEN INTEGRITY

- ○ Specimen intact
- ○ Fragmented specimen
- ○ Cannot be assessed, *specify*

TUMOUR DIMENSIONS

Maximum tumour dimension (largest tumour)

mm

Additional dimensions (largest tumour)

mm x mm

- ○ Cannot be assessed, *specify*

BLOCK IDENTIFICATION KEY

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

HISTOLOGICAL TUMOUR TYPE (select all that apply)

(Value list based on the World Health Organization Classification of Endocrine and Neuroendocrine Tumours, 5th Edition (2025))

- ☐ Phaeochromocytoma
- ☐ Extra-adrenal paraganglioma
- ☐ Composite phaeochromocytoma
  - ☐ Neuroblastoma, *specify* %
  - ☐ Ganglioneuroblastoma, *specify* %
  - ☐ Ganglioneuroma, *specify* %
  - ☐ Malignant peripheral nerve sheath tumour, *specify* %
- ☐ Composite paraganglioma
  - ☐ Neuroblastoma, *specify* %
  - ☐ Ganglioneuroblastoma, *specify* %
  - ☐ Ganglioneuroma, *specify* %
  - ☐ Malignant peripheral nerve sheath tumour, *specify* %
- ☐ Other, *specify* %

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TUMOUR NECROSIS

- ◯ Not identified
- ◯ Present

EXTENT OF INVASION (select all that apply)

- ◯ Cannot be assessed
- ◯ Not identified
- ☐ Microscopic transcapsular penetration of tumour capsule within an organ
- ☐ Microscopic transcapsular penetration of organ capsule
- ☐ Invasion into peritumoural soft tissue
- ☐ Invasion into adjacent structure(s)/organ(s), *specify*

LYMPHOVASCULAR INVASION

- ◯ Not identified
- ◯ Present

Type of vessel involved (select all that apply)

- ☐ Capillary
- ☐ Lymphatic
- ☐ Vein

Location of vessels (select all that apply)

- ☐ Periadrenal or peritumoral for extra-adrenal tumours, *specify*
  - ☐ Intracapsular ☐ Extracapsular
- ☐ Adrenal vein
- ☐ Vena cava
- ☐ Other (e.g., adrenal central vein and tributaries), *specify*

MARGIN STATUS

- ◯ Not involved (R0)

  Distance of tumour from closest margin ____ mm

  Specify closest margin(s) if possible
- ◯ Involved

  Extent
  - ◯ R1 (microscopic), *specify if possible*
  - ◯ R2 (macroscopic), *specify if possible*

  Location of involved margin(s), *specify if possible*
- ◯ Cannot be assessed, *specify*

PROLIFERATIVE FRACTION

Mitotic count ____ /2 mm²

AND/OR

Ki-67 proliferation index ____ %

- ◯ Cannot be assessed, *specify*

LYMPH NODE STATUS

- ◯ No nodes submitted or found
- ◯ Lymph node biopsy, *specify site(s) if applicable*

Number of lymph nodes examined ____

- ◯ Not involved
- ◯ Involved

  Number of involved lymph nodes ____
  - ◯ Number cannot be determined

ADVERSE HISTOLOGICAL FEATURES (select all that apply)

- ☐ Growth pattern
  - ◯ Large and irregular nests
  - ◯ Diffuse
  - ◯ Pseudorosette (even focal)
- ☐ Cellularity
  - ◯ Moderate (150–250 cells/U)
  - ◯ High (>250 cells/U)
- ☐ Cytologic features
  - ◯ Spindle cells
  - ◯ Other, *specify*
- ☐ Other, *specify*

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ANCILLARY STUDIES

- ◯ Not performed
- ◯ Performed (select all that apply)
  - ☐ Immunohistochemistry
    - ☐ Chromogranin A, *specify result(s)*
    - ☐ Keratins, *specify result(s)*
    - ☐ S100, *specify result(s)*
    - ☐ SDHB, *specify result(s)*
    - ☐ GATA-3, *specify result(s)*
    - ☐ Other, *specify*
  - ☐ Molecular testing, *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 (UICC TNM 9th edition)[b]

(Applicable only to pheochromocytoma and sympathetic paraganglioma; not applicable to head and neck paraganglioma)

TNM Descriptors (only if applicable) (select all that apply)

- ☐ m - multiple primary tumours
- ☐ r - recurrent
- ☐ y - post-therapy

Primary tumour (pT)

- ◯ TX[c] Primary tumour cannot be assessed
- ◯ T0 No evidence of primary tumour
- ◯ T1 Pheochromocytoma 5 cm or less in greatest dimension, no extra-adrenal invasion
- ◯ T2 Pheochromocytoma greater than 5 cm in greatest dimension, no extra-adrenal invasion
  Paraganglioma of any size, no local invasion
- ◯ T3 Tumour of any size with local invasion, into adjoining tissues or adjacent organs[d]

Regional lymph nodes (pN)

- ◯ NX[c] Regional lymph nodes cannot be assessed
- ◯ N0 No regional lymph node metastasis
- ◯ N1 Regional lymph node metastasis

[b] *Reproduced with permission. Source: UICC TNM Classification of Malignant Tumours, 9th Edition, eds by James Brierley, Meredith Giuliani, Brian O'Sullivan, Brian Rous, Elizabeth Van Eycken. 2025, Publisher Wiley (incorporating errata published 12th October 2025).*

[c] *TX and NX should be used only if absolutely necessary.*

[d] *Adjacent organs include kidney, liver, pancreas and spleen.*

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