

# Phaeochromocytoma and Paraganglioma Histopathology Reporting Guide



Family/Last name

Date of birth

Given name(s)

Patient identifiers

Date of request

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 (select all that apply)

- Information not provided
- Hormonal status
- 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
- Cannot be determined (testing status not known)

Imaging findings, *specify*

Relevant biopsy/cytology results, *specify*

Previous therapy (including pre-operative embolization, chemotherapy, radiotherapy, targeted therapy, immunotherapy), *specify*

Relevant familial history, *specify*

Presence of endocrine or other tumours, *specify*

Germline mutation or familial syndrome, *specify mutation if known*

Other, *specify*

## OPERATIVE PROCEDURE (select all that apply)

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

## SPECIMEN(S) SUBMITTED (select all that apply)

- Not specified
- Adrenal gland
  - Left       Right
- Biopsy tissue, *specify site(s) and laterality*
- Lymph nodes, *specify biopsy/dissection, site(s) and laterality*
- Other (e.g., right neck mass, midline abdominal mass), *specify site(s) and laterality*

## TUMOUR FOCALITY

- Unifocal
- Multiple
  - Multifocal (separate tumours in the same organ), *specify number of tumours*
  - Multiple tumours in separate organs,<sup>a</sup> *specify number of tumours*
- Indeterminate
- Cannot be assessed, *specify*

<sup>a</sup> If multiple tumours from different organs are present, separate datasets should be used to record all following elements for each tumour.

**TUMOUR SITE<sup>a</sup>** (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***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***MEDULLARY HYPERPLASIA** *(Applicable to adrenal specimens only)***Medullary nodules (microphaeochromocytoma) (<10 mm)** Present Absent Indeterminate Cannot be assessed, *specify***Diffuse hyperplasia** Present Absent Indeterminate Cannot be assessed, *specify***HISTOLOGICAL TUMOUR TYPE** (select all that apply) *(Value list from the World Health Organization Classification of Tumours: Pathology and Genetics of Tumours of Endocrine Organs (2017))* Pheochromocytoma Extra-adrenal paraganglioma Composite pheochromocytoma 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*⇒  %**EXTENT OF INVASION** (select all that apply)  Cannot be assessed 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 (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 to closest margin  mm
  - Closest margin, specify if possible
- Involved
  - Extent**
    - R1 (microscopic), specify if possible  mm
    - R2 (macroscopic), specify if possible  mm
  - Location of involved margin(s), specify if possible
- Cannot be assessed, specify

**PROLIFERATIVE FRACTION** 

- Mitotic count  /2 mm<sup>2</sup>
- AND/OR
- Ki-67  %
- Cannot be assessed

**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 positive lymph nodes
    - Number cannot be determined
- Extranodal extension (ENE)**
  - Not identified
  - Present
    - Number of nodes with ENE
  - Cannot be determined
- Maximum dimension of largest lymph node metastasis  mm

**ADVERSE FEATURES** (select all that apply) **Histological features**

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

**Other features**

- Extra-adrenal abdominal or mediastinal location
- Size >50 mm
- Negative staining for SDHB
- Biochemical testing showing high levels of methoxytyramine

**ANCILLARY STUDIES** (select all that apply) 

- Not performed
- Immunohistochemistry performed
  - Chromogranin A, specify result
  - Synaptophysin, specify result
  - S-100, specify result
  - SDHB, specify result
  - Tyrosine hydroxylase, specify result
  - Other, specify
- Molecular testing performed, specify result(s) if available
- Other, specify

**HISTOLOGICALLY CONFIRMED DISTANT METASTASES** 

- Not identified
- Not assessed
- Present, *specify site(s)*

**PATHOLOGICAL STAGING (AJCC TNM 8<sup>th</sup> edition)<sup>b</sup>** 

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

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

**Primary tumour (pT)**

- TX Primary tumour cannot be assessed
- T1 Pheochromocytoma <5 cm in greatest dimension, no extra-adrenal invasion
- T2 Pheochromocytoma ≥5 cm or paraganglioma-sympathetic of any size, no extra-adrenal invasion
- T3 Tumour of any size with invasion into surrounding tissues (e.g., liver, pancreas, spleen, kidneys)

Pheochromocytoma: within adrenal gland

Paraganglioma sympathetic: functional

Paraganglioma parasympathetic: often non-functional, and located in the head and neck

Note: Parasympathetic paragangliomas are not staged.

**Regional lymph nodes (pN)**

- NX Regional lymph nodes cannot be assessed
- N0 No regional lymph node metastasis
- N1 Regional lymph node metastasis

<sup>b</sup> Used with the permission of the American College of Surgeons, Chicago, Illinois. The original source for this information is the AJCC Cancer Staging Manual, Eighth Edition (2016) published by Springer Science+Business Media.