**Neoplasms of the Heart, Pericardium, and Great Vessels**

**Histopathology Reporting Guide**

<table>
<thead>
<tr>
<th>Family/Last name</th>
<th>Gender</th>
<th>Date of birth</th>
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<td>DD – MM – YYYY</td>
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<table>
<thead>
<tr>
<th>Given name(s)</th>
<th>Patient identifiers</th>
<th>Date of request</th>
<th>Accession/Laboratory number</th>
</tr>
</thead>
<tbody>
<tr>
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<td>DD – MM – YYYY</td>
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</tbody>
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Elements in **black text** are REQUIRED. Elements in **grey text** are RECOMMENDED.

### NEOADJUVANT THERAPY
- Information not provided
- Not administered
- Administered (describe)

### OPERATIVE PROCEDURE (Note 1)
- Not specified
- Resection
- Endovascular biopsy
- Image guided percutaneous biopsy
- Explantation
- Other (specify)

### SPECIMEN INTEGRITY (Note 2)  
(Applicable for resection and explant specimens only)
- Indeterminate
- Intact
- Disrupted (describe)

### TUMOUR SITE(S) (select all that apply) (Note 3)
- Right atrium
- Left atrium
- Right ventricle
- Left ventricle
- Ventricular septum
- Atrial septum
- Valve (specify)
- Great vessel (specify)
- Pericardium
- Other submitted specimens (specify)

### TUMOUR FOCALITY (Note 4)
- Indeterminate
- Unifocal
- Multifocal (specify the number of tumours in the specimen and their locations)

### MAXIMUM DIMENSION OF PRIMARY TUMOUR (Note 5)
(Applicable for resection and explant specimens only)
- mm
- Cannot be assessed

### BLOCK IDENTIFICATION KEY (Note 6)
(List overleaf or separately with an indication of the nature and origin of all tissue blocks)

### HISTOLOGICAL TUMOUR TYPE (Note 7)
(Value list from the World Health Organization Classification of Tumours of the Lung, Pleura, Thymus and Heart. Fourth edition (2015))

#### Heart

**BENIGN**
- Rhabdomyoma
- Myxoma
- Papillary fibroelastoma
- Haemangioma
- Fibroma
- Cystic tumour of the atrioventricular node
- Other (specify)

**MALIGNANT**
- Angiosarcoma
- Undifferentiated pleomorphic sarcoma
- Myxofibrosarcoma
- Other (specify)

**TUMOURS OF UNCERTAIN BEHAVIOUR**
- Inflammatory myofibroblastic tumour
- Paraganglioma
Pericardium
- Solitary fibrous tumour
- Germ cell tumour
- Angiosarcoma
- Other (specify)

Great vessels
- Angiosarcoma
- Intimal sarcoma subtype
- Leiomyosarcoma
- Other (specify)

HISTOLOGICAL GRADE (Note 8)
(Applicable to sarcomas only)
- Cannot be assessed
- Grade 1
- Grade 2
- Grade 3
- Ungraded sarcoma

RESPONSE TO NEOADJUVANT THERAPY (Note 11)
- Cannot be assessed
- Not identified
- Present

EXTENT OF INVASION (Note 10)
- Cannot be assessed
- No involvement of adjacent tissue(s)
- Involvement of adjacent tissue(s) (specify tissues)
- Other organ involvement (specify)

LYMPHOVASCULAR INVASION (Note 12)
(Applicable to solitary fibrous and germ cell tumours of the pericardium)
- Indeterminate
- Not identified
- Present
  Method of evaluation
  - Routine staining (H&E)
  - Immunohistochemistry for lymphovascular endothelium (specify)

ANCILLARY STUDIES (Note 13)
- Not performed
- Performed

IMMUNOHISTOCHEMISTRY (List stains)

MOLECULAR PATHOLOGY (List test(s))

CYTOGENETICS (List test(s))

OTHER (specify)

RESIDUAL VIGILANT TUMOUR
- Residual viable tumour %

MARGIN STATUS
- Not applicable (biopsies only)
- Cannot be assessed
- Not involved
- Involved (specify margin(s))
Note 1 - Operative procedure (Required)

Reason/Evidentiary Support

Because there may be more than one approach or technique to removing or sampling a tumour at a given location within the heart, specifying the nature of the operative procedure to the extent possible is important.¹

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Note 2 – Specimen integrity (Recommended)

Reason/Evidentiary Support

This element applies only to resection and explant specimens. If the tumour specimen is not received whole and intact, specify the nature of disruption (removed piecemeal, rupture during removal, etc.) This element has relevance to completeness of tumour removal and suitability for staging and size comparison with imaging studies.

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Note 3 – Tumour site (Required)

Reason/Evidentiary Support

The tumour site within the heart has implications in terms of obstruction of blood flow, valvular dysfunction and downstream vascular beds at risk of embolization and haematogenous spread.¹

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Note 4 – Tumour focality (Required)

Reason/Evidentiary Support

Multiple tumours may be present at the same site (e.g. left atrium in Carney Syndrome) or at different sites. A single tumour may invade multiple structures and thereby also be present in multiple cardiac locations. The tumour focality element clarifies this issue.

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Note 5 – Maximum dimension of primary tumour (Required)

Reason/Evidentiary Support

This element applies only to resection and explant specimens in which the entire tumour can be measured. Reporting the size in biopsy and other incomplete tumour samples may be misleading clinically.

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Note 6 – Block identification key (Recommended)

Reason/Evidentiary Support

The origin/designation of all tissue blocks should be recorded. This information should be documented in the final pathology report and is particularly important should the need for internal or external review arise. The reviewer needs to be clear about the origin of each block in order to provide an informed specialist opinion. If this information is not included in the final pathology report, it should be available on the laboratory computer system and relayed to the reviewing pathologist.

Recording the origin/designation of tissue blocks also facilitates retrieval of blocks for further immunohistochemical or molecular analysis, research studies or clinical trials.
Note 7 – Histological tumour type (Required)

Reason/Evidentiary Support

There are a large number of additional tumours that may occur in the heart, pericardium, and great vessels.\(^2\) Only the more common entities are specifically mentioned in this element, but other types should be entered under "other" (a complete list of histological types of primary tumours of the heart is included below). The neoplastic nature of some mass-forming lesions (lipomatous hypertrophy of the atrial septum, vascular malformations, hamartoma of mature cardiac myocytes, histiocytoid cardiomyopathy, etc.) may be in doubt.\(^1,3\) Whether or not to require a dataset worksheet on these masses is left to the discretion of the pathologist. (Note: for pericardial mesotheliomas, please use the thoracic dataset for pleural mesothelioma; haematolymphoid tumours are not covered by this dataset and will be dealt with in a future dataset).

This dataset is for tumours that arise primarily within the heart, pericardium, and great arteries. Metastatic lesions to these sites should not be recorded using this dataset.

### WHO classification of tumours of the heart\(^a,b\)

<table>
<thead>
<tr>
<th>Descriptor</th>
<th>ICD0 codes</th>
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<tbody>
<tr>
<td><strong>Benign tumours and tumour-like lesions</strong></td>
<td></td>
<td><strong>Malignant tumours</strong></td>
<td></td>
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<tr>
<td>Rhabdomyoma</td>
<td>8900/0</td>
<td>Angiosarcoma</td>
<td>9120/3</td>
</tr>
<tr>
<td>Histiocytoid cardiomyopathy</td>
<td></td>
<td>Undifferentiated pleomorphic sarcoma</td>
<td>8830/3</td>
</tr>
<tr>
<td>Hamartoma of mature cardiac myocytes</td>
<td></td>
<td>Osteosarcoma</td>
<td>9180/3</td>
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<tr>
<td>Adult cellular rhabdomyoma</td>
<td>8904/0</td>
<td>Myxofibrosarcoma</td>
<td>8811/3</td>
</tr>
<tr>
<td>Cardiac myxoma</td>
<td>8840/0</td>
<td>Leiomyosarcoma</td>
<td>8890/3</td>
</tr>
<tr>
<td>Papillary fibroelastoma</td>
<td>9120/0</td>
<td>Rhabdomyosarcoma</td>
<td>8900/3</td>
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<tr>
<td>Haemangioma, NOS</td>
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<td>Miscellaneous sarcomas</td>
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<tr>
<td>Capillary haemangioma</td>
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<tr>
<td>Cavernous haemangioma</td>
<td>9121/0</td>
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<td>Cardiac fibroma</td>
<td>8810/0</td>
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<tr>
<td>Lipoma</td>
<td>8850/0</td>
<td></td>
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<tr>
<td>Cystic tumour of the atrioventricular node</td>
<td>8454/0</td>
<td>Tumours of the pericardium</td>
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<tr>
<td>Granular cell tumour</td>
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<td>Solitary fibrous tumour</td>
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<td>Schwannoma</td>
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<td>Malignant</td>
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<td></td>
<td>Angiosarcoma</td>
<td>9120/3</td>
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<tr>
<td><strong>Tumours of uncertain behaviour</strong></td>
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<td>Synovial sarcoma</td>
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<td>Inflammatory myofibroblastic tumour</td>
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<td>Malignant mesothelioma</td>
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<td>Paraganglioma</td>
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<td>Germ cell tumours</td>
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<td></td>
<td>Teratoma, mature</td>
<td>9080/0</td>
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<tr>
<td><strong>Germ cell tumours</strong></td>
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<td>Teratoma, immature</td>
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<td>Mixed germ cell tumour</td>
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<tr>
<td>Teratoma, immature</td>
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<tr>
<td>Yolk sac tumour</td>
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</table>

\(^a\) The morphology codes are from the International Classification of Diseases for Oncology (ICD-O). Behaviour is coded /0 for benign tumours; /1 for unspecified, borderline, or uncertain behaviour; /2 for carcinoma in situ and grade III intraepithelial neoplasia; and /3 for malignant tumours.

\(^b\) The classification is modified from the previous WHO classification, taking into account changes in our understanding of these lesions.

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Note 8 – Histological grade\(^{1,2,4,5}\) (Required)

**Reason/Evidentiary Support**

This element only applies to malignant sarcomas of the heart, pericardium, and great vessels. This element captures information shown to be prognostically important in malignant sarcomas at other body sites. Evidence that these have the same importance in sarcomas of the heart, pericardium and great vessels is lacking.

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Note 9 – Mitotic count

**Reason/Evidentiary Support**

Mitotic figure count should be expressed as 
"#/mm\(^2\)" owing to the fact that differing field diameters of high power (x40) objectives dramatically vary the size of a single high power field (hpf). For example the hpf area for an x40 objective with a 0.40 mm field diameter is 0.125 mm\(^2\) whereas for an x40 objective with a 0.69 mm field diameter, the hpf area is 0.374 mm\(^2\). Depending on the objective used, it could take as many as 8 (for the 0.40 mm field diameter lens) or as few as 3 (for the 0.69 mm field diameter lens) hpfs to cover 1 mm\(^2\) of tissue. Each pathologist should determine the number of hpfs in a mm\(^2\) based on the field diameter of their x40 objective.\(^6\)

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Note 10 – Extent of invasion (Required)

**Reason/Evidentiary Support**

For the purposes of this data element, the parietal pericardium represents the anatomic boundary between the heart tissues and adjacent organs. Tumours that extend beyond the parietal pericardium should be considered "other organ involvement". Tumours crossing tissue boundaries in the heart (e.g. one chamber to another, across a valve, or into the pericardium) should be considered "involvement of adjacent tissues".\(^1,2\)

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Note 11 – Response to neoadjuvant therapy (Recommended)

**Reason/Evidentiary Support**

This element is not required since it presupposes knowledge of treatment prior to tumour removal. It may not always be possible to separate spontaneous tumour necrosis from treatment related necrosis. As of yet, no established level of pathologic response to treatment has been associated with prognostic significance.\(^2\)

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Note 12 – Lymphovascular invasion\(^7\) (Recommended)

Reason/Evidentiary Support

This element is commonly reported for malignancies, however since the majority of tumours in the heart and great vessels exist within the vasculature and have immediate access to haematogenous dissemination. This element should only be reported for pericardial tumours, such as germ cell tumours and solitary fibrous tumour, that do not arise within the vascular system.

Increasingly, centres are utilizing immunohistochemistry for antigens such as CD34, CD31, and/or D2-40 (podoplanin) to assess lymphovascular invasion. This may have an effect prognostically, but further study is needed. This element is not required, but will help in providing evidence along this line.

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Note 13 – Ancillary studies\(^2\) (Recommended)

Reason/Evidentiary Support

Increasingly, ancillary studies are needed to confirm and clarify a diagnosis. There is also potential for these kinds of studies to identify a target for therapy or confer meaningful prognostic information.

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References


