Neoplasms of the Heart, Pericardium, and Great Vessels Histopathology Reporting Guide

Family/Last name

Given name(s)

Gender ☐ Male ☐ Female

Date of birth DD – MM – YYYY

Patient identifiers

Date of request DD – MM – YYYY

Accession/Laboratory number

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)*
- ○ Not specified

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 graded
- ◯ Grade 1
- ◯ Grade 2
- ◯ Grade 3
- ◯ Ungraded sarcoma

NECROSIS

- ◯ Cannot be assessed
- ◯ Not identified
- ◯ Present

Extent of necrosis ____ %

MITOTIC COUNT (Note 9)
(most proliferative area)

____ /mm^2

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)*

RESPONSE TO NEOADJUVANT THERAPY (Note 11)

- ◯ Cannot be assessed
- ◯ Not identified
- ◯ Present

Residual viable tumour ____ %

MARGIN STATUS

- ◯ Not applicable *(biopsies only)*
- ◯ Cannot be assessed
- ◯ Not involved
- ◯ Involved *(specify margin(s))*

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)*

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]

Back

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.

Back

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]

Back

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.

Back

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.

Back

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.

Back

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]

Descriptor	ICD0 codes	Descriptor	ICD0 codes
Benign tumours and tumour-like lesions		**Malignant tumours**	
Rhabdomyoma	8900/0	Angiosarcoma	9120/3
Histiocytoid cardiomyopathy		Undifferentiated pleomorphic sarcoma	8830/3
Hamartoma of mature cardiac myocytes		Osteosarcoma	9180/3
Adult cellular rhabdomyoma	8904/0	Myxofibrosarcoma	8811/3
Cardiac myxoma	8840/0	Leiomyosarcoma	8890/3
Papillary fibroelastoma		Rhabdomyosarcoma	8900/3
Haemangioma, NOS	9120/0	Synovial sarcoma	9040/3
Capillary haemangioma	9131/0	Miscellaneous sarcomas	
Cavernous haemangioma	9121/0	Cardiac lymphomas	
Cardiac fibroma	8810/0	Metastatic tumours	
Lipoma	8850/0		
Cystic tumour of the atrioventricular node	8454/0	**Tumours of the pericardium**	
Granular cell tumour	9580/0	Solitary fibrous tumour	8815/1
Schwannoma	9560/0	Malignant	8815/3
		Angiosarcoma	9120/3
Tumours of uncertain behaviour		Synovial sarcoma	9040/3
Inflammatory myofibroblastic tumour	8825/1	Malignant mesothelioma	9050/3
Paraganglioma	8680/1	Germ cell tumours	
		Teratoma, mature	9080/0
Germ cell tumours		Teratoma, immature	9080/3
Teratoma, mature	9080/0	Mixed germ cell tumour	9085/3
Teratoma, immature	9080/3		
Yolk sac tumour	9071/3		

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.

Back

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.

Back

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]

Back

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]

Back

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]

Back

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.

Back

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.

Back

References

1 Burke AP, Tavora F, Maleszewski J and Frazier A (2015). *Tumors of the Heart and Great Vessels. AFIP Atlas of Tumor Pathology, Series 4*. ARP Press.

2 WHO (World Health Organization) (2015). *WHO Classification of Tumours of the Lung, Pleura, Thymus and Heart. Fourth edition* Travis WD, Brambilla E, Burke AP, Marx A and Nicholson AG. IARC Press, Lyon, France.

3 Thiene G (2013). Chapter 2 : cardiac tumours: classification and epidemiology. In: *Cardiac Tumor Pathology*, Basso C, Valente M and Thiene G (eds), Humana Press Inc.

4 Trojani M, Contesso G, Coindre JM, Rouesse J, Bui NB, de Mascarel A, Goussot JF, David M, Bonichon F and Lagarde C (1984). Soft-tissue sarcomas of adults; study of pathological prognostic variables and definition of a histopathological grading system. *Int J Cancer* 33:37-42.

5 Tazelaar HD, Locke TJ and McGregor CG (1992). Pathology of surgically excised primary cardiac tumors. *Mayo Clin Proc* 67(10):957-965.

6 NHS Cancer Screening Programmes and The Royal College of Pathologists (2005). *Pathology Reporting of Breast Disease. A Joint Document Incorporating the Third Edition of the NHS Breast Screening Programme's Guidelines for Pathology Reporting in Breast Cancer Screening and the Second Edition of The Royal College of Pathologists' Minimum Dataset for Breast Cancer Histopathology. NHSBSP Publication No 58.* http://www.cancerscreening.nhs.uk/breastscreen/publications/nhsbsp58-low-resolution.pdf.

7 Rose AE, Christos PJ, Lackaye D, Shapiro RL, Berman R, Mazumdar M, Kamino H, Osman I and Darvishian F (2011). Clinical relevance of detection of lymphovascular invasion in primary melanoma using endothelial markers D2-40 and CD34. *Am J Surg Pathol* 35(10):1441-1449.