

## **Protocol for the Examination of Specimens from Patients with Primary Malignant Tumors of the Heart**

**Protocol applies to primary malignant cardiac tumors. This protocol does not apply to hematolymphoid neoplasms or metastatic lesions involving the heart.**

### **Based on:**

CAP Cancer Protocol version 3.0.0.0.  
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### **Revision History:**

None

### **Summary of Changes:**

This protocol was retired by CAP on June 30, 2017.

### **Procedures Covered in this Protocol:**

- Excisional biopsy
- Partial resection
- Heart transplant

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**AAPA Macroscopic Examination Guidelines:  
Utilization of the CAP Cancer Protocols at the Surgical Gross Bench**

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The purpose of the Protocols is to support Laboratory Personnel engaged in the macroscopic examination of cancer resection specimens. The Protocols are based on specified relevant source documents, drafted by pathologists' assistant experts, and supported by information provided by the College of American Pathologists (CAP) and the American Joint Committee on Cancer (AJCC). These Protocols are intended to serve patients by ensuring that the macroscopic examination of cancer resection specimens is compliant with CAP Cancer Protocols, the AJCC Cancer Staging Manual, and provide optimization of the pre-analytic steps necessary to promote appropriate molecular studies.

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**Molecular Considerations:**

Cytogenetic studies are often useful in differentiating soft tissue sarcomas but are limited with cardiac primaries.

A reciprocal translocation t(X;18)(p11.2;q11.2) is seen in more than 90% of soft tissue synovial sarcomas and is specific for synovial sarcomas. This translocation can be detected by reverse transcriptase-polymerase chain reaction (RT-PCR).

**Specimen Handling Requirements:**

**RT-PCR** can be applied to paraffin-embedded sarcoma tissue, though fresh or frozen tissue is the preferred source. Small fragments of fresh tissue should be transported in RPMI media or RNA/later, and frozen tissue should be snap frozen in liquid nitrogen to preserve RNA & DNA. Frozen tissue should be stored at -70 degrees C. **Any tissues treated with decalcifying agents are not eligible for DNA analysis.**

**Reference laboratories** may have a preference for the type of tissue sent, depending on the methods used to isolate DNA. Consult the laboratory before tissue collection.

## **PROCEDURES AND GENERAL ANATOMIC CONSIDERATIONS:**

- **Procedures Covered by this Protocol: \***

- Excisional Biopsy
- Partial resection
- Heart transplant

\*This protocol does not apply to endomyocardial biopsies.

- **Specimen Size and Extent of Resection:**

- Weight (for all specimens)

- **Excisional biopsies and partial resection specimens:**

- Identification and three dimensions of specimen
  - Atrium, ventricle, interventricular septum
  - Measure myocardial thickness
- Identification of attached structures
  - Pericardium
  - Valves
    - ❖ Measure circumference of annulus
  - Coronary arteries
  - Papillary muscles
  - Thymus
  - Vena cava
  - Other (specify)

- **Native Heart**

- Identification and measurements of structures of the heart
  - Atria
    - ❖ Measure myocardial thickness.
    - ❖ Describe size of chamber, if complete – dilated, normal, restricted.
  - Ventricles
    - ❖ Measure myocardial thickness 1-2 cm below valves.
    - ❖ Describe size of chamber, if complete – dilated, normal, restricted.
  - Interventricular septum
    - ❖ Measure myocardial thickness 1-2 cm below valves.
  - Valves
    - ❖ Measure circumference of annulus.
    - ❖ Coronary arteries
    - ❖ Grade of stenosis: mild, moderate, or severe.

- **Specimen Integrity:**

State if specimen is intact, disrupted or indeterminate. Identify and describe any defects or disruptions. Consultation with the surgeon for clarification should be considered, especially in cardiac explant specimens.

## TUMOR ("T" of TNM)

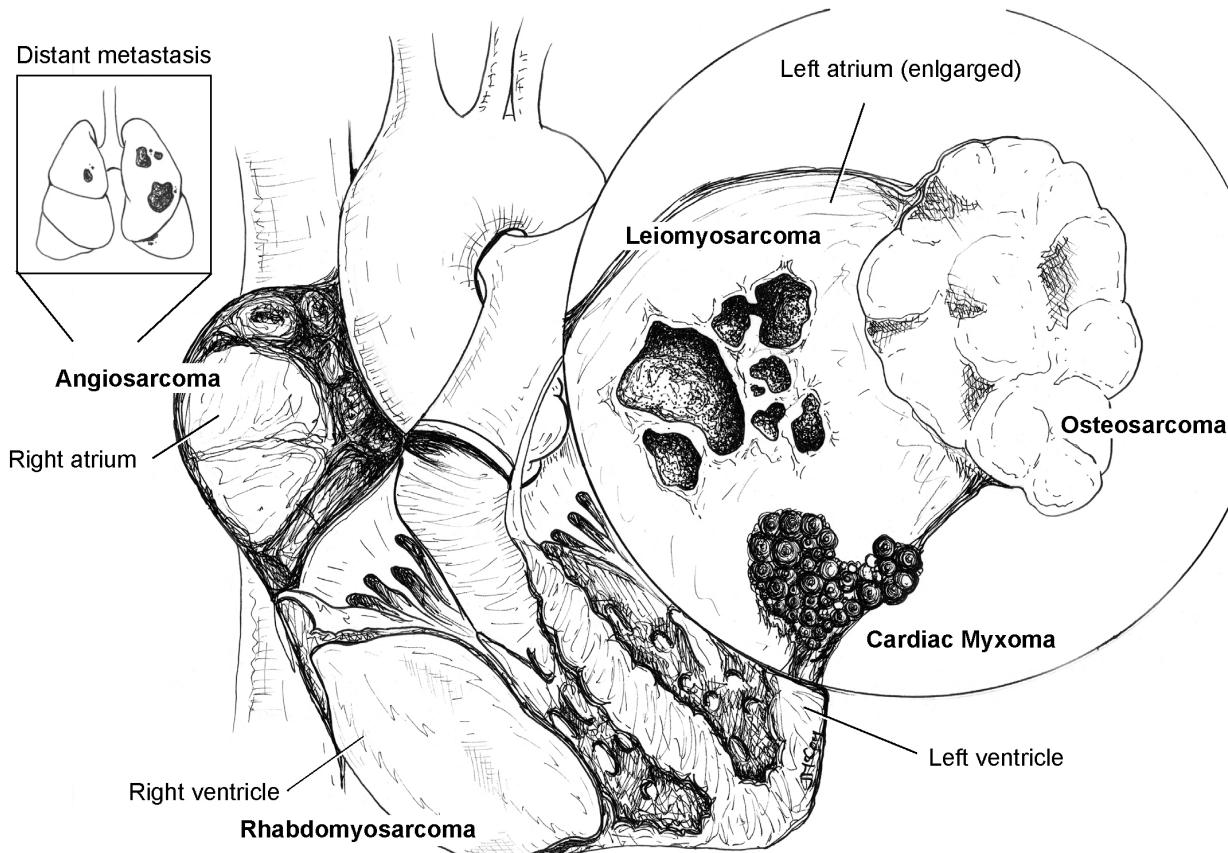
- **Tumor Size:**

- Include three dimensions for each tumor identified.

- **Tumor Site(s):**

- Pericardium
- Right ventricle
- Left ventricle
- Right atrium
- Left atrium
- Interventricular septum
- Other (specify)

Most malignant primary tumors of the heart are sarcomas. Angiosarcomas most often arise in the right atrium (80%), but often metastasize before a diagnosis is made. Therefore, they are often inoperable. Other sarcomas, such as osteosarcomas and leiomyosarcomas, tend to occur in the left side of the heart, and may be confused clinically with benign lesions like cardiac myxomas, which most often arise in the left atrium. These lesions may be surgically excised under the clinical suspicion of myxoma and turn out to be a sarcoma. Another lesion that may be resected under the clinical suspicion of sarcoma but turns out to be benign are mural thrombi. Points of attachment should be noted.



■ **Tumor Depth of Invasion and Relationship to Attached Organs / Structures:** \*

A published TNM staging system does not exist for primary cardiac tumors. Prognosis is indicated by the extent of involvement of surrounding tissues. Diffuse involvement has a poor prognosis.

Indicate involvement of adjacent tissues (if present):

- Valve leaflets/cusps
- Papillary muscles
- Aorta
- Pulmonary artery
- Pulmonary veins
- Pericardium
- Mediastinal tissue
- Superior vena cava
- Inferior vena cava

Indicate other organ involvement or lack of other organ involvement:

- Lung
- Pleura

\* *The most significant prognostic factors are location of tumor, extent of invasion into surrounding tissues, metastatic spread and extent of tumor necrosis (without treatment). A better prognosis is associated with tumor origin in the left atrium, as well as the absence of necrosis and metastasis, and a low mitotic count.*

■ **Margins:**

- The surgical margins should be differentiated from endocardial/epicardial surfaces and anatomically named if possible (interventricular septum margin vs. left ventricle margin).
  - Provide the distance of the tumor to all margins.
- If the margin is **macroscopically negative** for tumor:
  - The entire margin should be perpendicularly submitted, due to myocardial encroachment of tumor cells. Sarcoma cells often microscopically extend beyond leading edge of the grossly obvious tumor.
- If the margin is **macroscopically positive** for tumor:
  - Submit one representative section of the positive margin and give anatomic location.

Complete excision of sarcomas cannot be achieved in most cases, but partial resection may still provide months of symptom-free survival. Heart transplants may offer relatively long-term survival if there are no distant metastases.

■ **Explanatory Notes:**

Special considerations:

- Treatment effect:

The response to chemotherapy or radiation treatment should be reported with a positive or negative comment about the percentage of residual viable tumor.

One section per centimeter should be submitted of the residual viable tumor to allow accurate estimation of the percentage of residual viable tumor.

- Calcifications:

The presence of dense calcifications can indicate an osteosarcoma. Scattered or speckled calcifications often indicate benign lesions such as fibromas or myxomas. A well-fixed, decalcified section of these areas should be submitted.

- Necrosis:

The presence of necrosis is important for histologic grading. The current grading system of the Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) gives three scores:

Score 0: No necrosis

Score 1: <50% necrosis

Score 2: ≥50% necrosis

Sections should be well-fixed and cover a large representative area of the tumor for accurate grading.

**LYMPH NODES ("N" of TNM)**

■ **Lymph Nodes:**

Not applicable; a published TNM staging system for primary cardiac tumors does not exist.

## **METASTASIS ("M" of TNM)**

- **Metastasis:**

A published TNM staging system for primary cardiac tumors does not exist.

Metastasis indicates a poor prognosis. The most common metastatic sites, in order of descending frequency, are: lungs, regional lymph nodes, central nervous system, gastrointestinal tract, kidney, adrenal gland, thyroid, ovary, bone and pancreas.

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