SKIN

23

Carcinoma of the Skin (Excluding Eyelid, Vulva, and Penis)

C44.0 Skin of lip, NOS
C44.2 External ear
C44.3 Skin of other and unspecified parts of face
C44.4 Skin of scalp and neck
C44.5 Skin of trunk
C44.6 Skin of upper limb and shoulder
C44.7 Skin of lower limb and hip
C44.8 Overlapping lesion
C44.9 Skin, NOS

C63.2 Scrotum

This chapter applies to carcinomas of the skin, predominantly squamous cell carcinomas and basal cell carcinomas. Skin cancers are relatively common and for the most part have a good prognosis. Basal cell carcinomas, the most common cancers in humans, are easily treated with surgery. Staging of skin cancer depends on the size of the primary tumor. Refer to Chapter 38 for lesions on the eyelid and Chapter 24 for melanomas.

ANATOMY

Primary Site. The skin has two layers: an outer epidermis and the inner dermis. The epidermis consists predominantly of stratified squamous epithelium, the external layer of which is keratinized. The dermis contains connective tissue and elastic fibers. Immediately below the dermis is the subcutaneous tissue. The sebaceous glands and other glands of the skin are found in the dermis and adjacent subcutaneous tissue. All skin components—epidermis, dermis, and adnexal structures—can give rise to malignant neoplasms.

Cancers can arise from any area of the skin. They are most common on those surfaces exposed to sunlight, including the face, ears, hands, and scalp. Cancers can also arise on the truncal regions and on the extremities. Basal cell carcinomas often occur on the face.

This classification also includes tumors arising in the anal margin.

Regional Lymph Nodes. The regional lymph nodes are those appropriate to the location of the primary tumor.

Unilateral Tumors

Head, neck	Ipsilateral preauricular, submandibular, cervical, and supraclavicular lymph nodes
Thorax	Ipsilateral axillary lymph nodes
Arm	Ipsilateral epitrochlear and axillary lymph nodes
Abdomen, loins and buttocks	Ipsilateral inguinal lymph nodes
Leg	Ipsilateral popliteal and inguinal lymph nodes
Anal margin and perianal skin	Ipsilateral inguinal lymph nodes

With tumors in the boundary zones between the above, the lymph nodes pertaining to the regions on both sides of the boundary zone are considered to be regional lymph nodes. The following 4 cm-wide bands are considered boundary zones:

Between (Right/left)	*Along* (Midline)
Head and neck/thorax	Claviculo-acromion-upper shoulder blade edge
Thorax/arm	Shoulder-axilla-shoulder
Thorax/abdomen, loins, buttocks	Front: Middle between navel and costal arch Back: Lower border of thoracic vertebrae (midtransverse-axis)
Abdomen, loins, and buttock/leg	Groin-trochanter-gluteal sulcus

For tumors arising on the leg, the iliac nodes are considered sites of distant metastasis and should be coded as M1.

Metastatic Sites. The most common metastatic site is the lung, especially for the squamous cell carcinomas. Other sites of distant spread are unusual. Basal cell carcinomas tend to erode locally, although rarely they may metastasize.

RULES FOR CLASSIFICATION

The clinical and pathologic classifications are identical. However, pathologic staging uses the symbol "p" as a prefix.

The classification applies only to carcinomas. There should be microscopic verification of the disease to permit division of cases by histologic type. The following are suggested procedures for assessing the T, N, and M categories.

Clinical Staging. Assessment of skin cancer is based on inspection and palpation of the involved area and the regional lymph nodes. Imaging studies of the underlying bony structures is important, especially in the scalp, if the lesion is fixed.

T categories: Clinical examination
N categories: Clinical examination
M categories: Examination and imaging

Pathologic Staging. Complete resection of the entire site is indicated. Confirmation of lymph node involvement is also required.

DEFINITION OF TNM

Definitions for clinical (cTNM) and pathologic (pTNM) classifications are the same.

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
Tis Carcinoma *in situ*
T1 Tumor 2 cm or less in greatest dimension
T2 Tumor more than 2 cm but not more than 5 cm in greatest dimension
T3 Tumor more than 5 cm in greatest dimension
T4 Tumor invades deep extradermal structures (e.g., cartilage, skeletal muscle, or bone)

Note: In the case of multiple simultaneous tumors, the tumor with the highest T category will be classified and the number of separate tumors will be indicated in parentheses; e.g., T2 (5).

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

STAGE GROUPING

Stage 0	Tis	N0	M0
Stage I	T1	N0	M0
Stage II	T2	N0	M0
	T3	N0	M0
Stage III	T4	N0	M0
	Any T	N1	M0
Stage IV	Any T	Any N	M1

HISTOPATHOLOGIC TYPE

The staging system is used primarily for squamous cell and basal cell carcinomas of the skin. It also applies to adenocarcinomas developing from sweat or sebaceous glands and a spindle cell variant of squamous cell carcinoma. Squamous cell tumors may also be described as verrucous.

A form of *in situ* carcinoma or intraepidermal carcinoma is often referred to as Bowen's disease. This lesion should be coded as Tis.

HISTOPATHOLOGIC GRADE (G)

GX Grade cannot be assessed
G1 Well differentiated
G2 Moderately differentiated
G3 Poorly differentiated
G4 Undifferentiated

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5. Safai B, Good RA: Basal cell carcinoma with metastasis: Review of literature. Arch Pathol Lab Med 101:327–331, 1977
6. Strayer DS, Santa Crus DJ: Carcinoma *in situ* of the skin: A review of histopathology. J Cutan Pathol 7:244–259, 1980

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age ____ Sex ____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

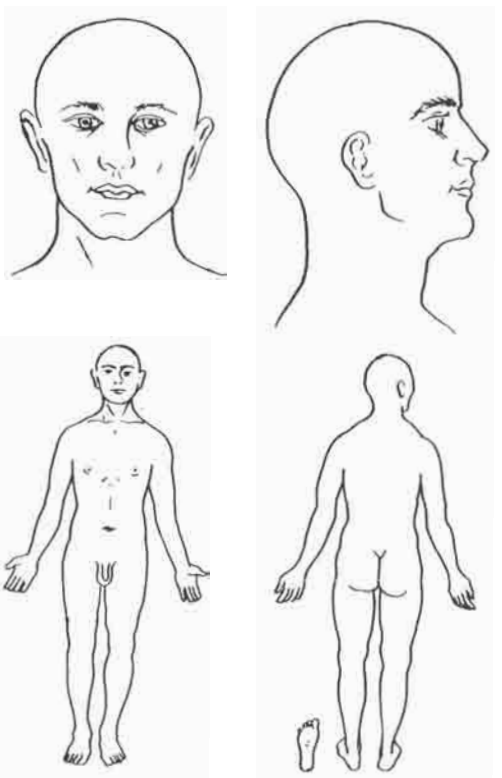
Clin	Path	DEFINITIONS	
		Primary Tumor (T)	
[ ]	[ ]	TX	Primary tumor cannot be assessed
[ ]	[ ]	T0	No evidence of primary tumor
[ ]	[ ]	Tis	Carcinoma *in situ*
[ ]	[ ]	T1	**Tumor 2 cm or less** in greatest dimension
[ ]	[ ]	T2	Tumor more than 2 cm but not more than 5 cm in greatest dimension
[ ]	[ ]	T3	Tumor more than 5 cm in greatest dimension
[ ]	[ ]	T4	Tumor invades deep extradermal structures, *i.e.*, cartilage, skeletal muscle or bone
		Lymph Node (N)	
[ ]	[ ]	NX	Regional lymph nodes cannot be assessed
[ ]	[ ]	N0	No regional lymph node metastasis
[ ]	[ ]	N1	Regional lymph node metastasis
		Distant Metastasis (M)	
[ ]	[ ]	MX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0	No distant metastasis
[ ]	[ ]	M1	Distant metastasis

Clin	Path	Stage Grouping			
[ ]	[ ]	0	Tis	N0	M0
[ ]	[ ]	I	T1	N0	M0
[ ]	[ ]	II	T2	N0	M0
			T3	N0	M0
[ ]	[ ]	III	T4	N0	M0
			Any T	N1	M0
[ ]	[ ]	IV	Any T	Any N	M1

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated
[ ] G3 Poorly differentiated
[ ] G4 Undifferentiated

Illustrations



Indicate on diagram primary tumor and regional nodes involved.

Histopathologic Type

The staging system is used primarily for squamous cell and basal cell carcinomas of the skin. It also applies to adenocarcinomas developing from sweat or sebaceous glands and a spindle cell variant of squamous cell carcinoma. Squamous cell tumors may also be described as verrucous.

A form of *in situ* carcinoma or intraepidermal carcinoma is oftren referred to as Bowen's disease. This lesion should be coded as Tis.

Staged by _____ M.D.
_____ Registrar
Date _____

24

Malignant Melanoma of the Skin (Excluding Eyelid)

C44.0 Skin of lip, NOS
C44.2 External ear
C44.3 Skin of other and unspecified parts of face
C44.4 Skin of scalp and neck
C44.5 Skin of trunk
C44.6 Skin of upper limb and shoulder
C44.7 Skin of lower limb and hip
C44.8 Overlapping lesion of skin
C44.9 Skin, NOS

C51 Vulva

C60 Penis

C63.2 Scrotum

Malignant melanomas are most common in fair-skinned persons, often with a history of chronic sun exposure. They can occur in any skin area, including the palms, soles, and nail beds. Rarely, melanomas may arise in other sites, such as the mucous membranes of the oral cavity, nasopharynx, vagina, urethra, and anal canal. Melanomas may also arise from the pigmented tissues of the eye and from giant hairy nevi. In some cases of disseminated disease, a primary lesion may not be found. Melanomas can be transmitted from mother to infant during pregnancy. Early detection and treatment of incipient melanomas have resulted in a significant decrease in the mortality from this disease. The staging classification outlined in this chapter applies only to melanomas arising in the skin. These tumors are staged histologically by measuring the depth of penetration into the underlying dermis or subcutis and by a statement on the level of invasion, with the cutaneous anatomic structures used as reference.

ANATOMY

Primary Site. The great majority of melanomas arise from the pigmented melanocytes located in the basal layer of the epidermis. The tumor often develops from a pre-existing pigmented lesion, although some arise from apparently normal skin. Melanomas are found on all skin surfaces. The tumor may grow into the dermis (nodular type) or spread horizontally along the skin (superficial spreading type). Multiple primary tumors may occur.

Regional Lymph Nodes. The regional lymph nodes depend on the location of the primary tumor. Regional nodes are as follows:

Unilateral Tumors

Head and neck	Ipsilateral preauricular, submandibular, cervical, and supraclavicular lymph nodes
Thorax	Ipsilateral axillary lymph nodes
Arm	Ipsilateral epitrochlear and axillary lymph nodes
Abdomen, loins, and buttocks	Ipsilateral inguinal lymph nodes
Leg	Ipsilateral popliteal and inguinal lymph nodes
Anal margin and perianal skin	Ipsilateral inguinal lymph nodes

With tumors in the boundary zones between the above, the lymph nodes pertaining to the regions on both sides of the boundary zone are considered regional lymph nodes. The following 4 cm-wide bands are considered as boundary zones:

Between (Right/left)	*Along* (Midline)
Head and neck/thorax	Claviculo-acromion-upper shoulder blade edge
Thorax/arm	Shoulder-axilla-shoulder
Thorax/abdomen, loins, and buttocks	Front: Middle between navel and costal arch Back: Lower border of thoracic vertebrae (midtransverse-axis)
Abdomen, loins, and buttock/leg	Groin-trochanter-gluteal sulcus

Iliac nodes are considered sites of distant metastasis and should be coded as M1.

Lesions arising in the midtransverse axis of the trunk at a level between the umbilicus and the lower costal margin anteriorly and extending laterally to the posterior level between the tenth thoracic spine (T10) and the first lumbar spine (L1) may spread with equal propensity to either contralateral or ipsilateral (or both) axillary or inguinal nodes.

Metastatic Sites. Melanomas can metastasize widely. No organ or tissue is exempt. In some cases, metastatic deposits may not become apparent for years. Melanomas commonly involve skin, subcutaneous tissues, lymph nodes, liver, bone, lung, brain, and visceral organs.

For staging purposes, two sub-M categories, identified as "a" and "b", are included. Metastasis to the skin, subcutaneous tissue, or lymph nodes beyond the site of the primary lymph node drainage is considered M1a. Metastasis to other distant sites—often referred to as visceral metastasis—is considered M1b. This distinction is based on the more favorable response to therapy by patients with skin or subcutaneous metastases only.

RULES FOR CLASSIFICATION

Clinical Staging. Clinical T classification ordinarily is not possible. Excisional biopsy and pathologic interpretation of the primary lesion are necessary for proper staging. Ulceration of the primary lesion may indicate a bad prognosis and should be recorded, but its presence does not alter staging.

Pathologic Staging. Pathologic staging of the primary melanoma is based on microscopic assessment of the depth of invasion and thickness of the primary tumor. Therefore, evaluation of the entire primary tumor, rather than a wedge or punch biopsy, is always advised. The entire thickness of the skin is needed for accurate classification. Regional nodes should be carefully evaluated, if available, and the number of positive nodes should be identified with the total number of lymph nodes removed.

Both the thickness and the level of invasion have prognostic significance, and both parameters should be reported by the pathologist.

Maximal thickness of the tumor is measured with an ocular micrometer at a right angle to the adjacent normal skin. The upper reference point is the top of the granular cell layer of the epidermis of the overlying skin, or the base of the lesion if the tumor is ulcerated. The lower reference point is usually the deepest point of invasion. It may be the invading edge of a single tumor mass or an isolated cell or group of cells deep to the main mass. Actual measurement should be recorded.

If no primary lesion is found, the tumor is coded as TX.

SURVIVAL ACCORDING TO AJCC STAGE
MELANOMA

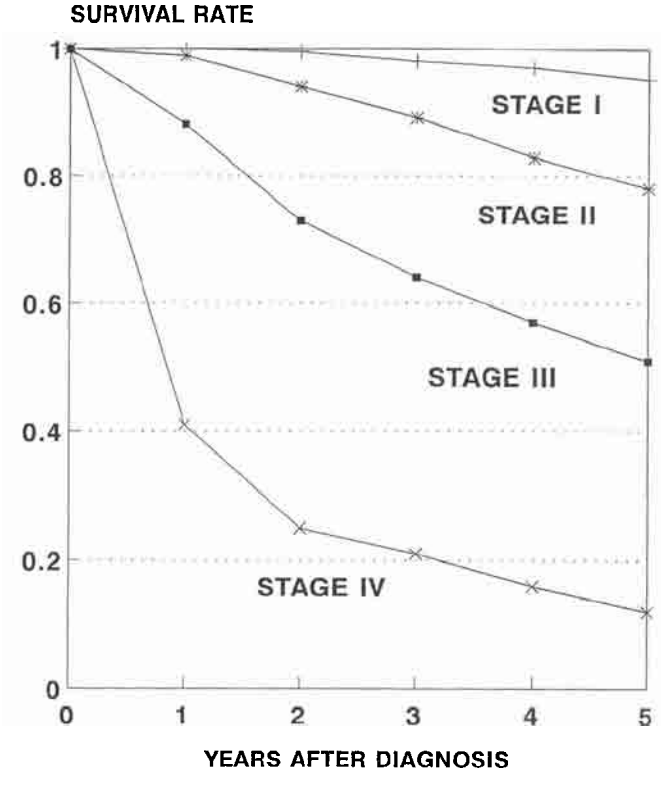


Fig. 24-1. Relative survival rates according to the stage of disease. Data taken from 8,479 patients who were diagnosed between 1977 and 1982. Patients are listed in the Surveillance, Epidemiology, and End Results Program of the National Cancer Institute. Stage I represents 4,286 patients; Stage II, 3,328; Stage III, 649; and Stage IV, 216.

DEFINITION OF TNM

Both the level of invasion and the maximum thickness determine the T classification and should be recorded. In case of discrepancy between tumor thickness and level, the pT category is based on the less favorable finding.

Satellite lesions or subcutaneous nodules more than 2 cm from the primary tumor but not beyond the site of the primary lymph node drainage are considered in-transit metastases and are listed under the N categories.

The extent of tumor is classified after excision.

Primary Tumor (pT)

- pTX Primary tumor cannot be assessed
- pT0 No evidence of primary tumor
- pTis Melanoma *in situ* (atypical melanocytic hyperplasia, severe melanocytic dysplasia), not an invasive lesion (Clark's Level I)
- pT1 Tumor 0.75 mm or less in thickness and invading the papillary dermis (Clark's Level II)
- pT2 Tumor more than 0.75 mm but not more than 1.5 mm in thickness and/or invades to the papillary-reticular dermal interface (Clark's Level III)
- pT3 Tumor more than 1.5 mm but not more than 4 mm in thickness and/or invades the reticular dermis (Clark's Level IV)
  - pT3a Tumor more than 1.5 mm but not more than 3mm in thickness
  - pT3b Tumor more than 3 mm but not more than 4 mm in thickness
- pT4 Tumor more than 4 mm in thickness and/or invades the subcutaneous tissue (Clark's Level V) and/or satellite(s) within 2 cm of the primary tumor
  - pT4a Tumor more than 4 mm in thickness and/or invades the subcutaneous tissue
  - pT4b Satellite(s) within 2 cm of the primary tumor

Regional Lymph Nodes (N)

- NX Regional lymph nodes cannot be assessed
- N0 No regional lymph node metastasis
- N1 Metastasis 3 cm or less in greatest dimension in any regional lymph node(s)
- N2 Metastasis more than 3 cm in greatest dimension in any regional lymph node(s) and/or in-transit metastasis
  - N2a Metastasis more than 3 cm in greatest dimension in any regional lymph nodes
  - N2b In-transit metastasis
  - N2c Both (N2a and N2b)

Distant Metastasis (M)

- MX Presence of distant metastasis cannot be assessed
- M0 No distant metastasis
- M1 Distant metastasis
  - M1a Metastasis in skin or subcutaneous tissue or lymph node(s) beyond the regional lymph nodes
  - M1b Visceral metastasis

Note: In-transit metastasis involves skin or subcutaneous tissue more than 2 cm from the primary tumor not beyond the regional lymph nodes.

STAGE GROUPING

Stage 0	pTis	N0	M0
Stage I	pT1	N0	M0
	pT2	N0	M0
Stage II	pT3	N0	M0
Stage III	pT4	N0	M0
	Any pT	N1	M0
	Any pT	N2	M0
Stage IV	Any pT	Any N	M1

HISTOPATHOLOGIC TYPE

The types of malignant melanoma are as follows:

Lentigo maligna (Hutchinson's freckle)
Radial spreading (superficial spreading)
Nodular
Acral lentiginous
Unclassified

A rare desmoplastic variant also exists.

Melanomas are identified according to site (e.g., mucosal, ocular, vaginal, anal, urethral). The staging classification described in this chapter applies only to those arising in the skin.

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5. Kopf AW, Rodriquez-Sains RS, Rigel DS, et al: "Small" melanomas: Relation of prognostic variables to diameter of superficial spreading melanomas. J Dermatol Surg Oncol 8:765–770, 1982

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	DEFINITIONS	
		Primary Tumor (pT)	
[ ]	[ ]	pTX	Primary tumor cannot be assessed
[ ]	[ ]	pT0	No evidence of primary tumor
[ ]	[ ]	pTis	Melanoma *in situ* (atypical melanocytic hyperplasia, severe melanocytic dysplasia, not an invasive lesion (Clark's Level I)
[ ]	[ ]	pT1	Tumor 0.75 mm or less in thickness and invades the papillary dermis (Clark's Level II)
[ ]	[ ]	pT2	Tumor more than 0.75 mm but not more than 1.5 mm in thickness and/or invades to papillary-reticular dermal interface (Clark's Level III)
[ ]	[ ]	pT3	Tumor more than 1.5 mm but not more than 4 mm in thickness and/or invades the reticular dermis (Clark's Level IV)
[ ]	[ ]	pT3a	Tumor more than 1.5 mm but not more than 3 mm in thickness
[ ]	[ ]	pT3b	Tumor more than 3 mm but not more than 4 mm in thickness
[ ]	[ ]	pT4	Tumor more than 4 mm in thickness and/or invades the subcutaneous tissue (Clark's Level V) and/or satellite(s) within 2 cm of the primary tumor
[ ]	[ ]	pT4a	Tumor more than 4 mm in thickness and/or invades the subcutaneous tissue
[ ]	[ ]	pT4b	Satellite(s) within 2 cm of primary tumor
		Lymph Node (N)	
[ ]	[ ]	NX	Regional lymph nodes cannot be assessed
[ ]	[ ]	N0	No regional lymph node metastasis
[ ]	[ ]	N1	Metastasis 3 cm or less in greatest dimension in any regional lymph node(s)
[ ]	[ ]	N2	Metastasis more than 3 cm in greatest dimension in any regional lymph node(s) and/or in-transit metastasis
[ ]	[ ]	N2a	Metastasis more than 3 cm in greatest dimension in any regional lymph node(s)
[ ]	[ ]	N2b	In-transit metastasis
[ ]	[ ]	N2c	Both (N2a and N2b)
		Distant Metastasis (M)	
[ ]	[ ]	MX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0	No distant metastasis
[ ]	[ ]	M1	Distant metastasis
[ ]	[ ]	M1a	Metastasis in skin or subcutaneous tissue or lymph node(s) beyond the regional lymph nodes
[ ]	[ ]	M1b	Visceral metastasis

Clin	Path	Stage Grouping			
[ ]	[ ]	I	pT1	N0	M0
			pT2	N0	M0
[ ]	[ ]	II	pT3	N0	M0
[ ]	[ ]	III	pT4	N0	M0
			Any pT	N1	M0
			Any pT	N2	M0
[ ]	[ ]	IV	Any pT	Any N	M1

Stage 0 pTis N0 M0

Staged by _____ M.D.
_____ Registrar
Date _____

(continued on next page)

Histopathologic Type

The types of malignant melanoma are as follows:

Lentigo maligna (Hutchinson's freckle)
Radial spreading (superficial spreading)
Nodular
Acral lentiginous
Unclassified

A rare desmoplastic variant also exists.
Melanomas are identified according to site (mucosal, ocular, vaginal, anal, urethral, and so forth). The staging classification described in this chapter applies only to those arising in the skin.

Sites of Distant Metastasis

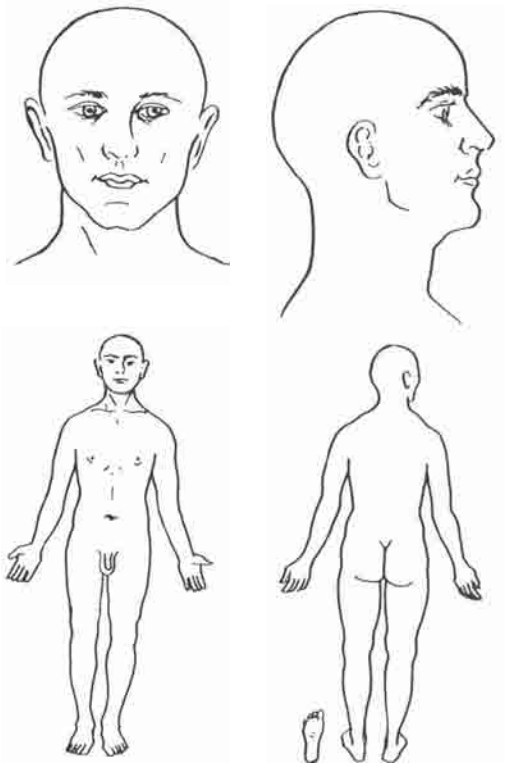
[ ] Pulmonary PUL
[ ] Osseous OSS
[ ] Hepatic HEP
[ ] Brain BRA
[ ] Lymph nodes LYM
[ ] Bone marrow MAR
[ ] Pleura PLE
[ ] Peritoneum PER
[ ] Skin SKI
[ ] Other OTH

Depth of Invasion

[ ] Level I (not a melanoma and further characterization is not necessary)
[ ] Level II
[ ] Level III
[ ] Level IV
[ ] Level V

Other description_____
Maximal thickness (mm)_____
Site of primary lesion (check diagram)_____
_____
Extent of primary lesion (include all pigmentation)_____
_____
Size in greatest diameter _____ cm

Illustrations



Indicate on diagrams primary tumor and regional nodes involved.

BREAST

25

Breast

C50.0 Nipple
C50.1 Central portion
C50.2 Upper-inner quadrant
C50.3 Lower-inner quadrant
C50.4 Upper-outer quadrant
C50.5 Lower-outer quadrant
C50.6 Axillary tail
C50.8 Overlapping lesion
C50.9 Breast, NOS

The following TNM definitions and stage groupings for carcinoma of the breast are the same for the AJCC and the UICC/TNM projects. This staging system for carcinoma of the breast applies to infiltrating and *in situ* carcinomas. Microscopic confirmation of the diagnosis is mandatory and the histologic type and grade of carcinoma should be recorded.

ANATOMY

Primary Site. Situated on the anterior chest wall, the mammary gland is composed of glandular tissue within a dense fibroareolar stroma. The glandular tissue consists of approximately 20 lobes, each of which terminates in a separate excretory duct in the nipple.

Regional Lymph Nodes. The breast lymphatics drain by way of three major routes: axillary, transpectoral, and internal mammary. Intramammary lymph nodes are considered with the axillary lymph nodes for staging purposes. Metastases to any other lymph nodes—including supraclavicular, cervical, and contralateral internal mammary nodes—are considered distant (M1). (Please refer to diagram.) The regional lymph nodes are:

(1) Axillary (ipsilateral): interpectoral (Rotter's) nodes and lymph nodes along the axillary vein and its tributaries, which may be divided into the following levels:

(i) Level I (low-axilla): lymph nodes lateral to the lateral border of the pectoralis minor muscle

(ii) Level II (mid-axilla): lymph nodes between the medial and lateral borders of the pectoralis minor muscle and the interpectoral (Rotter's) lymph nodes

(iii) Level III (apical axilla): lymph nodes medial to the medial margin of the pectoralis minor muscle, including those designated as subclavicular, infraclavicular, or apical.

Note: Intramammary lymph nodes are coded as axillary lymph nodes.

(2) Internal mammary (ipsilateral): lymph nodes in the intercostal spaces along the edge of the sternum in the endothoracic fascia.

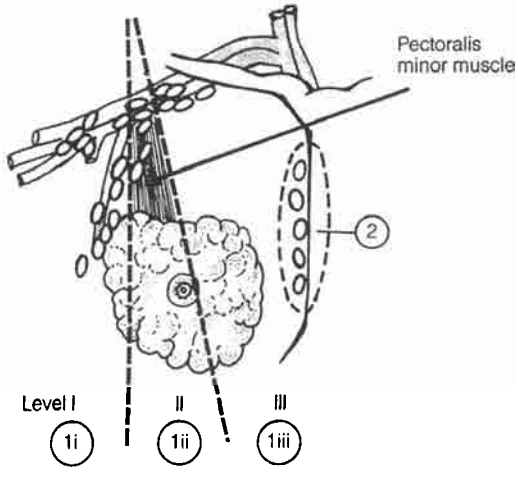
Any other lymph node metastasis is coded as a distant metastasis (M1), including supraclavicular, cervical, or contralateral internal mammary lymph nodes.

Metastatic Sites. All distant visceral sites are potential sites of metastases. The four major sites of involvement are bone, lung, brain, and liver, but this widely metastasizing disease has been found in almost every remote site.

RULES FOR CLASSIFICATION

Clinical Staging. Clinical staging includes physical examination, with careful inspection and palpation of

REGIONAL LYMPH NODES



the skin, mammary gland, and lymph nodes (axillary, supraclavicular, and cervical), pathologic examination of the breast or other tissues, and imaging to establish the diagnosis of breast carcinoma. The extent of tissues examined pathologically for clinical staging is less than that required for pathologic staging (see Pathologic Staging). Appropriate operative findings are elements of clinical staging, including the size of the primary tumor and chest wall invasion and the presence or absence of regional or distant metastasis.

Pathologic Staging. Pathologic staging includes all data used for clinical staging and surgical resection as well as pathologic examination of the primary carcinoma, including not less than excision of the primary carcinoma with no tumor in any margin of resection by gross pathologic examination. A case can be included in the pathologic stage if there is only microscopic, but not gross, involvement at the margin. If there is tumor in the margin of resection by gross examination, it is coded as TX, because the extent of primary tumor cannot be assessed. Resection of at least the low axillary lymph nodes (Level I)—that is, those lymph nodes located lateral to the lateral border of the pectoralis minor muscle—should be carried out. Such a resection ordinarily will include six or more lymph nodes. Metastatic nodules in the fat adjacent to the mammary carcinoma, without evidence of residual lymph node tissue, are considered regional lymph node metastases.

TNM CLASSIFICATION

Primary Tumor

The clinical measurement used for classifying the primary tumor (T) should be the one judged most accurate (e.g., physical examination or mammogram). Pathologically, the tumor size for classification (T) is a measurement of the invasive component. For example, if there is a large *in situ* component (4 cm) and a small invasive component (0.5 cm), the tumor is classified as T1a. The size of the primary tumor should be measured before any tissue is removed for special studies, such as for estrogen receptors.

Multiple Simultaneous Ipsilateral Primary Cancers

The following guidelines should be used when classifying multiple simultaneous ipsilateral primary (infiltrating, grossly measurable) carcinomas. These criteria do not apply to one grossly detected tumor associated with multiple separate microscopic foci.

1. Use the largest primary carcinoma to classify T.
2. Enter into the record that this is a case of multiple simultaneous ipsilateral primary carcinomas. Such cases should be analyzed separately.

Simultaneous Bilateral Breast Carcinomas

Each carcinoma is staged separately.

Inflammatory Carcinoma

Inflammatory carcinoma is a clinicopathologic entity characterized by diffuse brawny induration of the skin of the breast with an erysipeloid edge, usually without an underlying palpable mass. Radiologically, there may be a detectable mass and characteristic thickening of the skin over the breast. This clinical presentation is due to tumor embolization of dermal lymphatics. The tumor of inflammatory carcinoma is classified as T4d.

Paget's Disease of the Nipple

Paget's disease of the nipple without an associated tumor mass (clinical) or invasive carcinoma (pathologic) is classified as Tis. Paget's disease with a demonstrable mass (clinical) or an invasive component (pathologic) is classified according to the size of the tumor mass or invasive component.

Skin of the Breast

Dimpling of the skin, nipple retraction, or any other skin change except those described under T4b and T4d may occur in T1, T2, or T3 without changing the classification.

Chest Wall

The chest wall includes the ribs, intercostal muscles, and serratus anterior muscle but not the pectoral muscle.

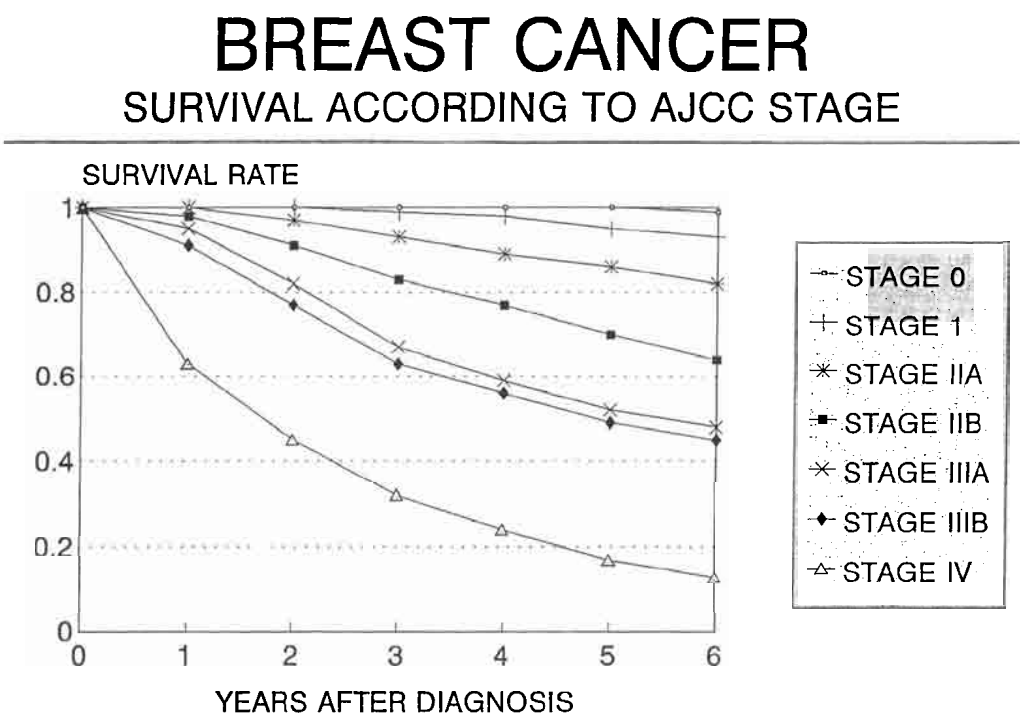


Fig. 25-1. Relative survival rates according to stage of disease. Data taken from 50,834 patients listed in the Surveillance, Epidemiology, and End Results Program of the National Cancer Institute. Patients were diagnosed between 1983 and 1987. Stage 0 represents 4,601 patients; Stage I, 16,519; Stage IIA, 14,692; Stage IIB, 8,283; Stage IIIA, 1,656; Stage IIIB, 1,389; and Stage IV, 3,694.

DEFINITION OF TNM

Primary Tumor (T)

Definitions for classifying the primary tumor (T) are the same for clinical and for pathologic classification. The telescoping method of classification can be applied. If the measurement is made by physical examination, the examiner will use the major headings (T1, T2, or T3). If other measurements, such as mammographic or pathologic, are used, the examiner can use the telescoped subsets of T1.

- TX Primary tumor cannot be assessed
- T0 No evidence of primary tumor
- Tis Carcinoma *in situ*: intraductal carcinoma, lobular carcinoma in situ, or Paget's disease of the nipple with no tumor
- T1 Tumor 2 cm or less in greatest dimension
  - T1a 0.5 cm or less in greatest dimension
  - T1b More than 0.5 cm but not more than 1 cm in greatest dimension
  - T1c More than 1 cm but not more than 2 cm in greatest dimension
- T2 Tumor more than 2 cm but not more than 5 cm in greatest dimension
- T3 Tumor more than 5 cm in greatest dimension
- T4 Tumor of any size with direct extension to chest wall or skin
  - T4a Extension to chest wall
  - T4b Edema (including peau d'orange) or ulceration of the skin of the breast or satellite skin nodules confined to the same breast
  - T4c Both (T4a and T4b)
  - T4d Inflammatory carcinoma (See the definition of inflammatory carcinoma in the introduction.)

Note: Paget's disease associated with a tumor is classified according to the size of the tumor.

Regional Lymph Nodes (N)

- NX Regional lymph nodes cannot be assessed (e.g., previously removed)
- N0 No regional lymph node metastasis
- N1 Metastasis to movable ipsilateral axillary lymph node(s)
- N2 Metastasis to ipsilateral axillary lymph node(s) fixed to one another or to other structures
- N3 Metastasis to ipsilateral internal mammary lymph node(s)

Pathologic Classification (pN)

pNX Regional lymph nodes cannot be assessed (e.g., previously removed, or not removed for pathologic study)
pN0 No regional lymph node metastasis
pN1 Metastasis to movable ipsilateral axillary lymph node(s)
- pN1a Only micrometastasis (none larger than 0.2 cm)
- pN1b Metastasis to lymph node(s), any larger than 0.2 cm
  - pN1bi Metastasis in one to three lymph nodes, any more than 0.2 cm and all less than 2 cm in greatest dimension
  - pN1bii Metastasis to four or more lymph nodes, any more than 0.2 cm and all less than 2 cm in greatest dimension
  - pN1biii Extension of tumor beyond the capsule of a lymph node metastasis less than 2 cm in greatest dimension
  - pN1biv Metastasis to a lymph node 2 cm or more in greatest dimension

pN2 Metastasis to ipsilateral axillary lymph nodes that are fixed to one another or to other structures
pN3 Metastasis to ipsilateral internal mammary lymph node(s)

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis (includes metastasis to ipsilateral supraclavicular lymph node(s))

STAGE GROUPING

Stage 0	Tis	N0	M0
Stage I	T1	N0	M0
Stage IIA	T0	N1	M0
	T1	N1*	M0
	T2	N0	M0
Stage IIB	T2	N1	M0
	T3	N0	M0
Stage IIIA	T0	N2	M0
	T1	N2	M0
	T2	N2	M0
	T3	N1	M0
	T3	N2	M0
Stage IIIB	T4	Any N	M0
	Any T	N3	M0
Stage IV	Any T	Any N	M1

* *Note:* The prognosis of patients with N1a is similar to that of patients with pN0.

HISTOPATHOLOGIC TYPE

The histologic types are as follows:

Carcinoma, NOS (not otherwise specified)
Ductal
- Intraductal (*in situ*)
- Invasive with predominant intraductal component
- Invasive, NOS
- Comedo
- Inflammatory
- Medullary with lymphocytic infiltrate
- Mucinous (colloid)
- Papillary
- Scirrhous
- Tubular
- Other

Lobular
- *In situ*
- Invasive with predominant *in situ* component
- Invasive

Nipple
- Paget's disease, NOS
- Paget's disease with intraductal carcinoma
- Paget's disease with invasive ductal carcinoma
- Other

Undifferentiated carcinoma

HISTOPATHOLOGIC GRADE (G)

GX Grade cannot be assessed
G1 Well differentiated
G2 Moderately differentiated
G3 Poorly differentiated
G4 Undifferentiated

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age ____ Sex ____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path		
		DEFINITIONS	
		Primary Tumor (T)	
[ ]	[ ]	TX	Primary tumor cannot be assessed
[ ]	[ ]	T0	No evidence of primary tumor
[ ]	[ ]	Tis	Carcinoma *in situ:* Intraductal carcinoma, lobular carcinoma *in situ*, or Paget's disease of the nipple with no tumor
[ ]	[ ]	T1	Tumor 2 cm or less in greatest dimension
[ ]	[ ]	T1a	0.5 cm or less in greatest dimension
[ ]	[ ]	T1b	More than 0.5 cm but not more than 1 cm in greatest dimension
[ ]	[ ]	T1c	More than 1 cm but not more than 2 cm in greatest dimension
[ ]	[ ]	T2	Tumor more than 2 cm but not more than 5 cm in greatest dimension
[ ]	[ ]	T3	Tumor more than 5 cm in greatest dimension
[ ]	[ ]	T4	Tumor of any size with direct extension to chest wall or skin
[ ]	[ ]	T4a	Extension to chest wall
[ ]	[ ]	T4b	**Edema (including peau** d'orange) or ulceration of the skin of breast or satellite skin nodules confined to same breast
[ ]	[ ]	T4c	Both T4a and T4b
[ ]	[ ]	T4d	Inflammatory carcinoma
		Lymph Node (N)	
[ ]	[ ]	NX	Regional lymph nodes cannot be assessed (*e.g.* previously removed)
[ ]	[ ]	N0	No regional lymph node metastasis
[ ]	[ ]	N1	Metastasis to movable ipsilateral axillary lymph node(s)
[ ]	[ ]	N2	Metastasis to ipsilateral axillary lymph node(s) fixed to one another or to other structures
[ ]	[ ]	N3	Metastasis to ipsilateral internal mammary lymph node(s)
		Pathologic Classification (pN)	
[ ]	[ ]	pNX	Regional lymph nodes cannot be assessed (*e.g.* previously removed, or not removed for pathologic study)
[ ]	[ ]	pN0	No regional lymph node metastasis
[ ]	[ ]	pN1	Metastasis to movable ipsilateral axillary lymph node(s)
[ ]	[ ]	pN1a	Only micrometastasis (none larger than 0.2 cm)
[ ]	[ ]	pN1b	Metastasis to lymph nodes, any larger than 0.2 cm
[ ]	[ ]	pN1bi	Metastasis in 1 to 3 lymph nodes, any more than 0.2 cm and all less than 2 cm in greatest dimension
[ ]	[ ]	pN1bii	Metastasis to 4 or more lymph nodes, any more than 0.2 cm and all less than 2 cm in greatest dimension
[ ]	[ ]	pN1biii	Extension of tumor beyond the capsule of a lymph node metastasis less than 2 cm in greatest dimension
[ ]	[ ]	pN1biv	Metastasis to a lymph node 2 cm or more in greatest dimension
[ ]	[ ]	pN2	Metastasis to ipsilateral axillary lymph nodes that are fixed to one another or to other structures
[ ]	[ ]	pN3	Metastasis to ipsilateral internal mammary lymph node(s)
		Distant Metastasis (M)	
[ ]	[ ]	MX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0	No distant metastasis
[ ]	[ ]	M1	Distant metastasis (includes metastasis to ipsilateral supraclavicular lymph node(s))

(continued on next page)

Clin	Path	Stage Grouping			
[ ]	[ ]	0	Tis	N0	M0
[ ]	[ ]	I	T1	N0	M0
[ ]	[ ]	IIA	T0	N1	M0
			T1	N1*	M0
			T2	N0	M0
[ ]	[ ]	IIB	T2	N1	M0
			T3	N0	M0
[ ]	[ ]	IIIA	T0	N2	M0
			T1	N2	M0
			T2	N2	M0
			T3	N1	M0
			T3	N2	M0
[ ]	[ ]	IIIB	T4	Any N	M0
			Any T	N3	M0
[ ]	[ ]	IV	Any T	Any N	M1

**Note: The prognosis of patients with pN1a is similar to that of patients with pN0.*

Staged by _____ M.D.

_____ Registrar

Date _____

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated
[ ] G3 Poorly differentiated
[ ] G4 Undifferentiated

Histopathologic Type

The histologic types are the following:

Carcinoma, NOS (not otherwise specified)
Ductal
- Intraductal (*in situ*)
- Invasive with predominant intraductal component
- Invasive, NOS (not otherwise specified)
- Comedo
- Inflammatory
- Medullary with lymphocytic infiltrate
- Mucinous (colloid)
- Papillary
- Scirrhous
- Tubular
- Other

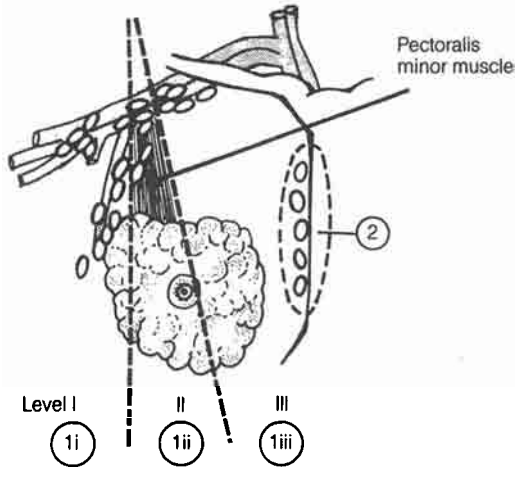
Lobular
- *In situ*
- Invasive with predominant *in situ* component
- Invasive

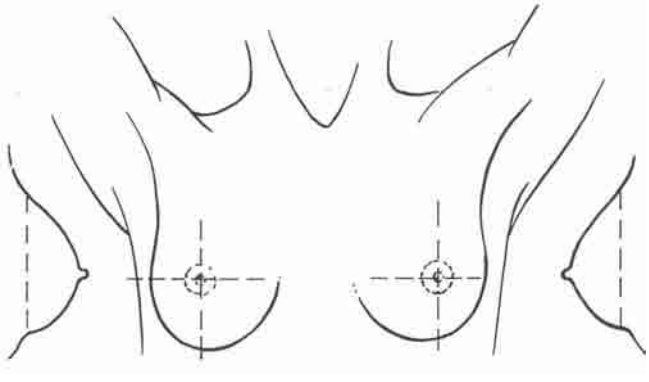
Nipple
- Paget's disease, NOS (not otherwise specified)
- Paget's disease with intraductal carcinoma
- Paget's disease with invasive ductal carcinoma

Other
- Undifferentiated carcinoma

Illustrations

REGIONAL LYMPH NODES





Indicate on diagram primary tumor and regional nodes involved.

GYNECOLOGIC TUMORS

The cervix uteri, corpus uteri, ovary, vagina, and vulva are the sites included in this section. The cervix uteri and corpus uteri were among the first sites to be classified by the TNM system. The League of Nations stages for carcinoma of the cervix have been used with minor modifications for nearly 50 years, and, because these are accepted by the International Federation of Gynecology and Obstetrics (FIGO), the TNM categories have been defined to correspond to the FIGO stages. Some amendments have been made in collaboration with FIGO, and the classifications now published have the approval of FIGO, AJCC, and all other national TNM committees of the Union Internationale Contre le Cancer (UICC).

The AJCC has worked closely with FIGO in the classification of cancer at gynecologic sites. Staging of malignant tumors is essentially the same, and stages are comparable in the two systems.

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Cervix Uteri

C53.0 Endocervix
C53.1 Exocervix
C53.8 Overlapping lesion
C53.9 Cervix uteri

ANATOMY

Primary Site. The cervix is the lower third of the uterus. It is roughly cylindrical in shape, projects through the upper anterior vaginal wall, and communicates with the vagina through an orifice called the external os. Cancer of the cervix may originate on the vaginal surface or in the canal.

Regional Lymph Nodes. The cervix is drained by preureteral, postureteral, and uterosacral routes into the following first station nodes:

Paracervical
Parametrial
Hypogastric (obturator)
Common iliac
Internal and external iliac
Presacral
Sacral

Para-aortic node involvement is considered distant metastasis and is coded as M1.

Metastatic Sites. The most common sites of distant spread are the lungs and skeleton.

RULES FOR CLASSIFICATION

The classification applies only to carcinomas. There should be histologic confirmation of the disease.

Clinical Staging. Careful clinical examination should be performed in all cases, preferably by an experienced examiner and with anesthesia. The clinical staging must not be changed because of subsequent findings. When doubt exists as to the stage to which a particular cancer should be allocated, the earlier stage is mandatory. The following examinations are permitted: palpation, inspection, colposcopy, endocervical curettage, hysteroscopy, cystoscopy, proctoscopy, intravenous urography, and x-ray examination of the lungs and skeleton. Suspected bladder or rectal involvement should be confirmed by biopsy and histologic evidence. Optional examinations include lymphangiography, arteriography, venography, laparoscopy, and other imaging methods. Because these are not yet generally available and because the interpretation of results is variable, the findings of optional studies should not be the basis for changing the clinical staging.

Pathologic Staging. In cases treated by surgical procedures, the pathologist's findings in the removed tissues

can be the basis for extremely accurate statements on the extent of disease. These findings should not be allowed to change the clinical staging but should be recorded in the manner described for the pathologic staging of disease. The pTNM nomenclature is appropriate for this purpose. Infrequently, hysterectomy is carried out in the presence of unsuspected extensive invasive cervical carcinoma. Such cases cannot be clinically staged or included in therapeutic statistics, but it is desirable that they be reported separately. Only if the rules for clinical staging are strictly observed will it be possible to compare results among clinics and by different modes of therapy.

Anatomic Subsites

Endocervix
Exocervix

DEFINITION OF TNM

The definitions of the T categories correspond to the several stages accepted by FIGO. Both systems are included for comparison.

Primary Tumor (T)

TNM	FIGO	DEFINITION
TX	—	Primary tumor cannot be assessed
T0	—	No evidence of primary tumor
Tis	—	Carcinoma *in situ*
T1	I	Cervical carcinoma confined to the uterus (extension to the corpus should be disregarded)
T1a	IA	Preclinical invasive carcinoma, diagnosed by microscopy only
T1a1	IA1	Minimal microscopic stromal invasion
T1a2	IA2	Tumor with an invasive component 5 mm or less in depth taken from the base of the epithelium and 7 mm or less in horizontal spread
T1b	IB	Tumor larger than T1a2
T2	II	Cervical carcinoma invades beyond the uterus but not to the pelvic wall or to the lower third of the vagina
T2a	IIA	Tumor without parametrial invasion
T2b	IIB	Tumor with parametrial invasion
T3	III	Cervical carcinoma extends to the pelvic wall and/or involves the lower third of the vagina and/or causes hydronephrosis or nonfunctioning kidney
T3a	IIIA	Tumor involves the lower third of the vagina, with no extension to the pelvic wall
T3b	IIIB	Tumor extends to the pelvic wall and/or causes hydronephrosis or a nonfunctioning kidney
T4*	IVA	Tumor invades the mucosa of the bladder or rectum and/ or extends beyond the true pelvis
M1	IVB	Distant metastasis

**Note:* Presence of bullous edema is not sufficient evidence to classify a tumor as T4.

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

TNM	FIGO	DEFINITION
MX		Presence of distant metastasis cannot be assessed
M0		No distant metastasis
M1	IVB	Distant metastasis

pTNM Pathologic Classification

The pT, pN, and pM categories correspond to the T, N, and M categories.

STAGE GROUPING

AJCC/UICC				FIGO
Stage 0	Tis	N0	M0	
Stage IA	T1a	N0	M0	Stage IA
Stage IB	T1b	N0	M0	Stage IB
Stage IIA	T2a	N0	M0	Stage IIA
Stage IIB	T2b	N0	M0	Stage IIB
Stage IIIA	T3a	N0	M0	Stage IIIA
Stage IIIB	T1	N1	M0	Stage IIIB
	T2	N1	M0	
	T3a	N1	M0	
	T3b	Any N	M0	
Stage IVA	T4	Any N	M0	Stage IVA
Stage IVB	Any T	Any N	M1	Stage IVB

HISTOPATHOLOGIC TYPE

Cases should be classified as carcinoma of the cervix if the primary growth is in the cervix. All histologic types must be included. Grading is encouraged but is not a basis for modifying the stage groupings. When surgery is the primary treatment, the histologic findings permit the case to have pathologic staging. In this, the pTNM nomenclature is to be used.

The histopathologic types are:

Cervical intraepithelial neoplasia, grade III
Squamous cell carcinoma *in situ*
Squamous cell carcinoma
  Keratinizing
  Nonkeratinizing
  Verrucous
Adenocarcinoma *in situ*
Adenocarcinoma *in situ*, endocervical type
Endometrioid adenocarcinoma
Clear cell adenocarcinoma
Adenosquamous carcinoma
Adenoid cystic carcinoma
Small cell carcinoma
Undifferentiated carcinoma

HISTOPATHOLOGIC GRADE (G)

GX Grade cannot be assessed
G1 Well differentiated
G2 Moderately differentiated
G3 Poorly differentiated
G4 Undifferentiated

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	TNM category	FIGO* stage	DEFINITIONS
				Primary Tumor (T)
[ ]	[ ]	TX		Primary tumor cannot be assessed
[ ]	[ ]	T0		No evidence of primary tumor
[ ]	[ ]	Tis		Carcinoma *in situ*
[ ]	[ ]	T1	I	**Cervical carcinoma** confined to uterus (extension to corpus should be disregarded)
[ ]	[ ]	T1a	IA	Preclinical invasive carcinoma, diagnosed by microscopy only
[ ]	[ ]	T1a1	IA1	Minimal microscopic stromal invasion
[ ]	[ ]	T1a2	IA2	**Tumor with invasive component 5 mm** or less in depth taken from the base of the epithelium and 7 mm or less in horizontal spread
[ ]	[ ]	T1b	IB	Tumor larger than T1a2
[ ]	[ ]	T2	II	Cervical carcinoma invades beyond uterus but not to pelvic wall or to the lower third of vagina
[ ]	[ ]	T2a	IIA	Tumor without parametrial invasion
[ ]	[ ]	T2b	IIB	Tumor with parametrial invasion
[ ]	[ ]	T3	III	Cervical carcinoma extends to pelvic wall and/or involves lower third of vagina and/or causes hydronephrosis or nonfunctioning kidney
[ ]	[ ]	T3a	IIIA	Tumor involves lower third of vagina, no extension to pelvic wall
[ ]	[ ]	T3b	IIIB	Tumor extends to pelvic wall and/or causes hydronephrosis or nonfunctioning kidney
[ ]	[ ]	T4**	IVA	Tumor invades mucosa of bladder or rectum and/or extends beyond the true pelvis
				Lymph Node (N)
[ ]	[ ]	NX		Regional lymph nodes cannot be assessed
[ ]	[ ]	N0		No regional lymph node metastasis
[ ]	[ ]	N1		Regional lymph node metastasis
				Distant Metastasis (M)
[ ]	[ ]	MX		Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0		No distant metastasis
[ ]	[ ]	M1	IVB	Distant metastasis

Stage Grouping

Clin	Path	AJCC/UICC				FIGO
[ ]	[ ]	0	Tis	N0	M0	
[ ]	[ ]	IA	T1a	N0	M0	I A
[ ]	[ ]	IB	T1b	N0	M0	I B
[ ]	[ ]	IIA	T2a	N0	M0	II A
[ ]	[ ]	IIB	T2b	N0	M0	II B
[ ]	[ ]	IIIA	T3a	N0	M0	III A
[ ]	[ ]	IIIB	T1	N1	M0	III B
			T2	N1	M0	
			T3a	N1	M0	
			T3b	Any N	M0	
[ ]	[ ]	IVA	T4	Any N	M0	IV A
[ ]	[ ]	IVB	Any T	Any N	M1	IV B

* FIGO: *Federation Internationale de Gynecologie et d'Obstetrique*
** Note: *Presence of bullous edema is not sufficient evidence to classify a tumor T4.*

Staged by _____ M.D.
_____ Registrar
Date _____

(continued on next page)

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated
[ ] G3 Poorly differentiated
[ ] G4 Undifferentiated

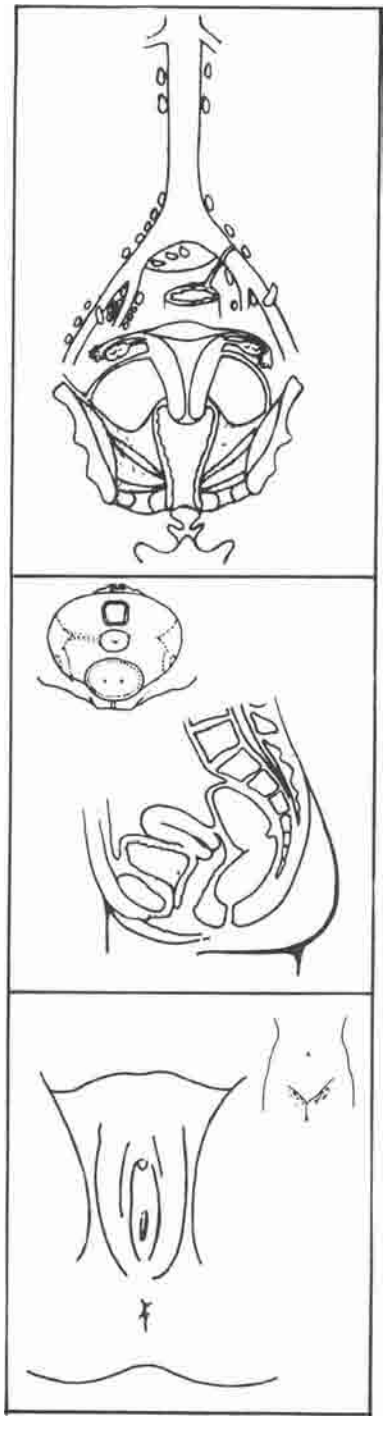
Histopathologic Type

Cases should be classified as carcinoma of the cervix if the primary growth is in the cervix. All histologic types must be included. Grading is encouraged but is not a basis for modifying the stage groupings. When surgery is the primary treatment, the histologic findings permit the case to have pathologic staging. In this, the pTNM nomenclature is to be used.

The histopathologic types are:

Cervical intraepithelial neoplasia, grade III
Squamous cell carcinoma *in situ*
Squamous cell carcinoma
  Keratinizing
  Nonkeratinizing
  Verrucous
Adenocarcinoma *in situ*
Adenocarcinoma *in situ*, endocervical type
Endometroid adenocarcinoma
Clear cell adenocarcinoma
Adenosquamous carcinoma
Adenoid cystic carcinoma
Small cell carcinoma
Undifferentiated carcinoma

Illustrations



Indicate on diagrams primary tumor and regional nodes involved.

27

Corpus Uteri

C54.0 Isthmus uteri
C54.1 Endometrium
C54.2 Myometrium
C54.3 Fundus uteri
C54.8 Overlapping lesion
C54.9 Corpus uteri
C55.9 Uterus, NOS

ANATOMY

Primary Site. The corpus uteri refers to the upper two-thirds of the uterus above the level of the internal cervical os. The fallopian tubes enter at the upper lateral corners of this pear-shaped body. The portion lying above a line joining the tubo-uterine orifices is often referred to as the fundus.

Regional Lymph Nodes. The regional lymph nodes include:

Para-aortic
Parametrial
Presacral
Internal iliac
Paracervical
Obturator
Hypogastric
External iliac
Common iliac
Presacral
Sacral promontory (Gerota's)
Uterosacral

Metastatic Sites. The vagina and lung are the common metastatic sites.

RULES FOR CLASSIFICATION

The classification applies only to carcinoma. There should be histologic verification and grading of the tumor.

Clinical Staging. Careful clinical staging should be performed, preferably by an experienced examiner and with anesthesia, before any definitive therapy begins. The clinical staging must not be changed because of subsequent findings. When doubt exist about the stage to which a particular cancer should be allocated, the earlier stage is mandatory. The following examinations are permitted: palpation, inspection, colposcopy, endocervical curettage, hysteroscopy, cystoscopy, proctoscopy, intravenous urography, and imaging examination of lungs and skeleton. Optional examinations include lymphangiography, arteriography, venography, and laparoscopy. Sounding and determination of uterine cavity depth is an important step. Fractional curettage is essential, with separation of endometrial and endocervical curettings. Careful inspection and palpation of the vagina should be done to assess the entire length of the vaginal tube from the apex to the urethra.

Pathologic Staging. Hysterectomy with or without pelvic node dissection provides the basis for surgical-pathologic staging and should not be substituted for clinical staging.

Anatomic Subsites

Corpus uteri
Isthmus uteri

DEFINITION OF TNM

The definitions of the T categories correspond to the several stages accepted by FIGO. (FIGO stages are fur-

ther subdivided by histologic grade of tumor.) Both systems are included for comparison.

Primary Tumor (T)

TNM	FIGO	DEFINITION
TX	—	Primary tumor cannot be assessed
T0	—	No evidence of primary tumor
Tis	—	Carcinoma in situ
T1	I	Tumor confined to the corpus uteri
T1a	IA	Tumor limited to the endometrium
T1b	IB	Tumor invades up to or less than one-half of the myometrium
T1c	IC	Tumor invades more than one-half of the myometrium
T2	II	Tumor invades the cervix but not extending beyond the uterus
T2a	IIA	Endocervical glandular involvement only
T2b	IIB	Cervical stromal invasion
T3 and/or N1	III	Local and/or regional spread as specified in T3a, b, N1 and FIGO IIIA, B, and C below
T3a	IIIA	Tumor involves the serosa and/or adnexa (direct extension or metastasis) and/or cancer cells in ascites or peritoneal washings
T3b	IIIB	Vaginal involvement (direct extension or metastasis)
N1	IIIC	Metastasis to the pelvic and/or paraaortic lymph nodes
T4*	IVA	Tumor invades the bladder mucosa or the rectum and/or the bowel mucosa
M1	IVB	Distant metastasis (*excluding* metastasis to the vagina, pelvic serosa, or adnexa; *including* metastasis to intra-abdominal lymph nodes other than paraaortic, and/or inguinal lymph nodes.)

* *Note:* The presence of bullous edema is not sufficient evidence to classify a tumor as T4.

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

TNM	FIGO	DEFINITION
MX	—	Presence of distant metastasis cannot be assessed
M0	—	No distant metastasis
M1	IVB	Distant metastasis

pTNM Pathologic Classification

The pT, pN, and pM categories correspond to the T, N, and M categories.

STAGE GROUPING

AJCC/UICC				FIGO
Stage 0	Tis	N0	M0	
Stage IA	T1a	N0	M0	Stage IA
Stage IB	T1b	N0	M0	Stage IB
Stage IC	T1c	N0	M0	Stage IC
Stage IIA	T2a	N0	M0	Stage IIA
Stage IIB	T2b	N0	M0	Stage IIB
Stage IIIA	T3a	N0	M0	Stage IIIA
Stage IIIB	T3b	N0	M0	Stage IIIB
Stage IIIC	T1	N1	M0	Stage IIIC
	T2	N1	M0	
	T3a	N1	M0	
	T3b	N1	M0	
Stage IVA	T4	Any N	M0	Stage IVA
Stage IVB	Any T	Any N	M1	Stage IVB

NOTES ABOUT STAGING

Studies of large series of cases of endometrial carcinoma limited to the corpus have shown that the prognosis is related to some extent to the size of the uterus. However, uterine enlargement may be caused by fibroids, adenomyosis, and other disorders. Therefore, the size of the uterus cannot serve as a basis for subgrouping Stage I cases. The length and the width of the uterine cavity are related to the prognosis. The great majority of cases of corpus cancer belong to Stage I. Extension of the carcinoma to the endocervix is confirmed by fractional curettage, hysterography, or hysteroscopy. Scraping the cervix should be the first step of the curettage; the specimens from the cervix should be examined separately. Occasionally, it may be difficult to decide whether the endocervix is involved by the cancer. In such cases, the simultaneous presence of normal cervical glands and cancer in the same section will give the final diagnosis.

Extension of the carcinoma outside the uterus should refer a case to Stage III or Stage IV.

The presence of metastases in the vagina or in the ovary permits allotment of a case to Stage III.

HISTOPATHOLOGIC TYPE

It is desirable that Stage I cases be subgrouped according to the degree of differentiation described on microscopic examination. The predominant lesion is adenocarcinoma, but all histologic types should be reported. However, choriocarcinomas, sarcomas, mixed mesodermal

tumors, and carcinosarcomas should be presented separately.

The histopathologic types are:

Endometrioid carcinoma
- Adenocarcinoma
- Adenocanthoma (adenocarcinoma with squamous metaplasia)
- Adenosquamous carcinoma (mixed adenocarcinoma and squamous cell carcinoma)

Mucinous adenocarcinoma
Serous adenocarcinoma
Clear cell adenocarcinoma
Squamous cell adenocarcinoma
Undifferentiated adenocarcinoma

HISTOPATHOLOGIC GRADE (G)

GX	Grade cannot be assessed
G1	Well differentiated
G2	Moderately differentiated
G3-4	Poorly differentiated or undifferentiated

Note: For details of FIGO histologic grading of endometrial carcinoma, please see the following publications:

FIGO: Annual report on the results of treatment of gynecological cancer. Int J Gynecol Obstet 28:189–193, 1989

FIGO: Changes in gynecologic cancer staging by the International Federation of Gynecology and Obstetrics. Am J Obstet Gynecol 162: 610–611, 1990.

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	TNM category	FIGO* stage	DEFINITIONS
				Primary Tumor (T)
[ ]	[ ]	TX		Primary tumor cannot be assessed
[ ]	[ ]	T0		No evidence of primary tumor
[ ]	[ ]	Tis		Carcinoma in situ
[ ]	[ ]	T1	I	Tumor confined to corpus uteri
[ ]	[ ]	T1a	IA	Tumor limited to endometrium
[ ]	[ ]	T1b	IB	**Tumor invades up to or less than** one half of the myometrium
[ ]	[ ]	T1c	IC	Tumor invades to more than one half of the myometrium
[ ]	[ ]	T2	II	Tumor invades cervix but does not extend beyond uterus
[ ]	[ ]	T2a	IIA	Endocervical glandular involvement only
[ ]	[ ]	T2b	IIB	Cervical stromal invasion
[ ]	[ ]	T3 &/or N1	III	Local and/or regional spread as specified in T3a, b, N1 and FIGO IIIA, B and C below
[ ]	[ ]	T3a	IIIA	Tumor involves serosa and/or adnexae (direct extension or metastasis) and/or cancer cells in ascites or peritoneal washings
[ ]	[ ]	T3b	IIIB	Vaginal involvement (direct extension or metastis) r metastasis
[ ]	[ ]	N1	IIIC	Metastasis to the pelvic and/or paraaortic lymph nodes
[ ]	[ ]	T4**	IVA	Tumor invades bladder mucosa and/or bowel mucosa
[ ]	[ ]	M1	IVB	**Distant metastasis. (Excluding metastasis to vagina,** pelvic serosa or adnexae. Including metastasis to intraabdominal lymph nodes other than paraaortic, and/or inguinal lymph nodes.)

Lymph Node (N)

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

Distant Metastasis (M)

[ ]	[ ]	MX Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0 No distant metastasis
[ ]	[ ]	M1 IVB Distant metastasis

Stage Grouping

Clin	Path	AJCC/UICC				FIGO
[ ]	[ ]	0	Tis	N0	M0	
[ ]	[ ]	IA	T1a	N0	M0	Stage IA
		IB	T1b	N0	M0	Stage IB
		IC	T1c	N0	M0	Stage IC
[ ]	[ ]	IIA	T2a	N0	M0	Stage IIA
		IIB	T2b	N0	M0	Stage IIB
[ ]	[ ]	IIIA	T3a	N0	M0	Stage IIIA
[ ]	[ ]	IIIB	T3b	N0	M0	Stage IIIB
[ ]	[ ]	IIIC	T1	N1	M0	Stage IIIC
			T2	N1	M0	
			T3a	N1	M0	
			T3b	N1	M0	
[ ]	[ ]	IVA	T4	Any N	M0	Stage IVA
[ ]	[ ]	IVB	Any T	Any N	M1	Stage IVB

* *FIGO: Federation Internationale de Gynecologie et d'Obstetrique*
** *Note: The presence of bullous edema is not sufficient evidence to classify a tumor T4.*

Staged by _____ M.D.
_____ Registrar
Date _____

(continued on next page)

Histopathologic Type

It is desirable that Stage I cases be subgrouped according to the degree of differentiation described on microscopic examination. The predominant lesion is adenocarcinoma, but all histologic types should be reported. However, choriocarcinomas, sarcomas, mixed mesodermal tumors, and carcinosarcomas should be presented separately.

The histopathologic types are:

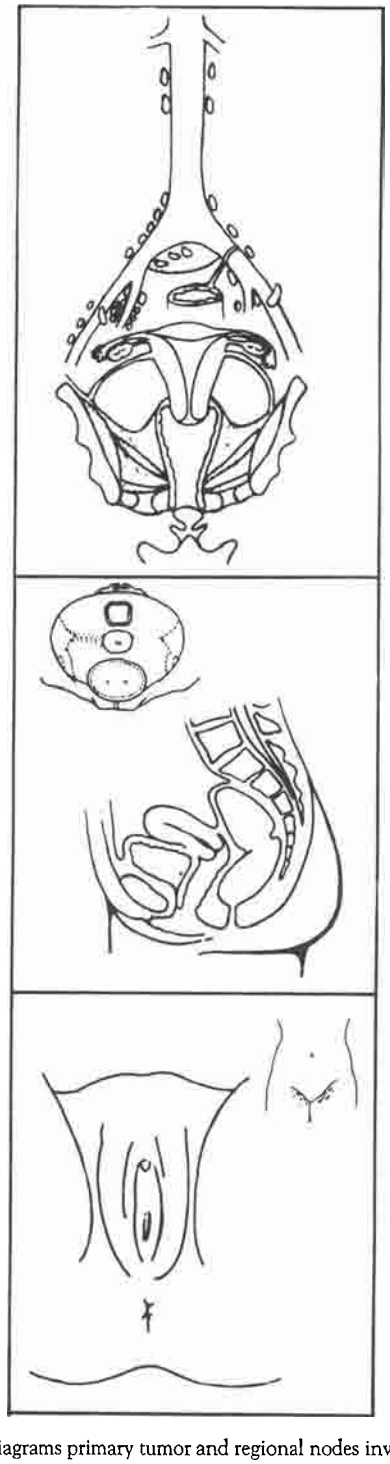
Endometrioid carcinoma
- Adenocarcinoma
- Adenocanthoma (adenocarcinoma with squamous metaplasia)
- Adenosquamous carcinoma (mixed adenocarcinoma and squamous cell carcinoma)

Mucinous adenocarcinoma
Serous adenocarcinoma
Clear cell adenocarcinoma
Squamous cell adenocarcinoma
Undifferentiated adenocarcinoma

Histopathologic Grade (G)

[ ]	GX	Grade cannot be assessed
[ ]	G1	Well differentiated
[ ]	G2	Moderately differentiated
[ ]	G3-G4	Poorly differentiated or undifferentiated

Illustrations



Indicate on diagrams primary tumor and regional nodes involved.

28

Ovary

C56.9 Ovary

ANATOMY

Primary Site. Ovaries are a pair of solid, flattened ovoids 2 to 4 cm in diameter connected by a peritoneal fold to the broad ligament and by the infundibulopelvic ligament to the lateral wall of the pelvis.

Regional Lymph Nodes. The lymphatic drainage occurs by the utero-ovarian and round ligament trunks and an external iliac accessory route into the following regional nodes:

External iliac
Common iliac
Hypogastric
Internal iliac
Obturator
Lateral sacral
Aortic nodes
Inguinal nodes
Pelvic, NOS
Retroperitoneal, NOS

Metastatic Sites. The peritoneum, including the omentum and pelvic and abdominal viscera, are common sites for seeding. Diaphragmatic involvement and liver metastases are common. Pulmonary and pleural involvement also occur.

RULES FOR CLASSIFICATION

There should be histologic confirmation of the disease to permit division of cases by histopathologic type. In accordance with FIGO,* a simplified version of the WHO** histologic typing (1973 publication No. 9) is recommended. The extent of differentiation (grade) should be recorded.

* FIGO: Federation Internationale de Gynecologie et d'Obstetrique
** World Health Organization

It is desirable to have a clinical stage grouping of ovarian tumors similar to those already existing for other malignant tumors in the female pelvis. Rarely is it possible to come to a final diagnosis by inspection or palpation or by any of the other methods recommended for clinical staging of carcinoma of the uterus and vagina. Therefore, the Cancer Committee of FIGO has recommended that clinical staging of primary carcinoma of the ovary be based on findings by laparoscopy or laparotomy, as well as on the usual clinical examination and roentgen studies.

Clinical Staging. Although clinical studies similar to those for other sites may be used, establishing a diagnosis most often requires a laparotomy, which is most widely accepted in clinical staging. Clinical studies include routine chest radiography. Computed tomography or other imaging studies may be helpful in both initial staging and follow-up of the tumors.

Pathologic Staging. This should include laparotomy and resection of ovarian masses, as well as hysterectomy. Biopsies of suspicious sites, such as the omentum, mesentery, liver, diaphragm, and pelvic and para-aortic nodes, are required.

DEFINITION OF TNM

The definitions of the T categories correspond to the several stages accepted by FIGO. Both systems are included for comparison.

Primary Tumor (T)

TNM	FIGO	DEFINITION
TX	—	Primary tumor cannot be assessed
T0	—	No evidence of primary tumor
T1	I	Tumor limited to ovaries (one or both)

TNM	FIGO	DEFINITION
T1a	IA	Tumor limited to one ovary; capsule intact, no tumor on ovarian surface, no malignant cells in ascites or peritoneal washings
T1b	IB	Tumor limited to both ovaries; capsules intact, no tumor on ovarian surface, no malignant cells in ascites or peritoneal washings
T1c	IC	Tumor limited to one or both ovaries with any of the following: capsule ruptured, tumor on ovarian surface, malignant cells in ascites or peritoneal washings
T2	II	Tumor involves one or both ovaries with pelvic extension
T2a	IIA	Extension and/or implants on the uterus and/or tube(s); no malignant cells in ascites or peritoneal washings
T2b	IIB	Extension to other pelvic tissues; no malignant cells in ascites or peritoneal washings
T2c	IIC	Pelvic extension (2a or 2b) with malignant cells in ascites or peritoneal washings
T3 and/or N1	III	Tumor involves one or both ovaries with microscopically confirmed peritoneal metastasis outside the pelvis and/or regional lymph node metastasis
T3a	IIIA	Microscopic peritoneal metastasis beyond the pelvis
T3b	IIIB	Macroscopic peritoneal metastasis beyond the pelvis 2 cm or less in the greatest dimension
T3c and/or N1	IIIC	Peritoneal metastasis beyond the pelvis more than 2 cm in the greatest dimension and/or regional lymph node metastasis
M1	IV	Distant metastasis (excludes peritoneal metastasis)

Note: Liver capsule metastasis is T3/Stage III; liver parenchymal metastasis, M1/Stage IV. Pleural effusion must have positive cytology for M1/Stage IV.

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

TNM	FIGO	DEFINITION
MX	—	Presence of distant metastasis cannot be assessed
M0	—	No distant metastasis
M1	IV	Distant metastasis (excludes peritoneal metastasis)

Note: The presence of nonmalignant ascites is not classified. The presence of ascites does not affect staging unless malignant cells are present.

pTNM Pathologic Classification

The pT, pN, and pM categories correspond to the T, N, and M categories.

STAGE GROUPING

AJCC/UICC				FIGO
Stage IA	T1a	N0	M0	Stage IA
Stage IB	T1b	N0	M0	Stage IB
Stage IC	T1c	N0	M0	Stage IC
Stage IIA	T2a	N0	M0	Stage IIA
Stage IIB	T2b	N0	M0	Stage IIB
Stage IIC	T2c	N0	M0	Stage IIC
Stage IIIA	T3a	N0	M0	Stage IIIA
Stage IIIB	T3b	N0	M0	Stage IIIB
Stage IIIC	T3c	N0	M0	Stage IIIC
	Any T	N1	M0	
Stage IV	Any T	Any N	M1	Stage IV

HISTOPATHOLOGIC TYPE

The types currently recommended are serous tumors, mucinous tumors, endometrioid tumors, clear cell (mesonephroid) tumors, undifferentiated tumors, and unclassified tumors. Malignant tumors other than those of the common epithelial types are not to be included with the categories listed below. However, the more common ones—such as granulosa cell tumor, immature teratoma, dysgerminoma, and endodermal sinus tumor—may be collected and reported separately by institutions so desiring, particularly those with a pediatric population among their patients.

The histopathologic types are:

Serous cystomas
- Serous cystadenomas with proliferating activity of the epithelial cells and nuclear abnormalities, but with no infiltrative destructive growth (low potential or borderline malignancy)
- Serous cystadenocarcinomas

Mucinous cystomas
- Mucinous cystadenomas with proliferating activity of the epithelial cells and nuclear abnormalities, but with no infiltrative destructive growth (low potential or borderline malignancy)
- Mucinous cystadenocarcinomas

Endometrioid tumors (similar to adenocarcinomas in the endometrium)
  Endometrioid tumors with proliferating activity of the epithelial cells and nuclear abnormalities, but with no infiltrative destructive growth (low potential or borderline malignancy)
  Endometrioid adenocarcinomas
Clear cell (mesonephroid) tumors
  Clear cell tumors with proliferating activity of the epithelial cells and nuclear abnormalities, but with no infiltrative destructive growth (low potential or borderline malignancy)
  Clear cell cystadenocarcinomas
Unclassified tumors

In some cases of inoperable widespread malignant tumor, it may be impossible for the gynecologist and the pathologist to decide the origin of the growth. In order to evaluate the results obtained in the treatment of carcinoma of the ovary, however, it is necessary that all patients be reported on, including those thought to have a malignant ovarian tumor. If clinical examination cannot exclude the possibility that the lesion is a primary ovarian carcinoma, the case should be reported in the group "special category" and belong to the histologic group of unclassified tumors.

HISTOPATHOLOGIC GRADE (G)

GX	Grade cannot be assessed
GB	Borderline malignancy
G1	Well differentiated
G2	Moderately differentiated
G3-4	Poorly differentiated or undifferentiated

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	TNM category	FIGO* stage	DEFINITIONS
				Primary Tumor (T)
[ ]	[ ]	TX		Primary tumor cannot be assessed
[ ]	[ ]	T0		No evidence of primary tumor
[ ]	[ ]	T1	I	Tumor limited to ovaries (one or both)
[ ]	[ ]	T1a	IA	Tumor limited to one ovary; capsule intact, no tumor on ovarian surface. No malignant cells in ascites or peritoneal washings.
[ ]	[ ]	T1b	IB	Tumor limited to both ovaries; capsules intact, no tumor on ovarian surface. No malignant cells in ascites or peritoneal washings.
[ ]	[ ]	T1c	IC	Tumor limited to one or both ovaries with any of the following: capsule ruptured, tumor on ovarian surface, malignant cells in ascites, or peritoneal washings.
[ ]	[ ]	T2	II	Tumor involves one or both ovaries with pelvic extension
[ ]	[ ]	T2a	IIA	Extension and/or implants on uterus and/or tube(s). No malignant cells in ascites or peritoneal washings.
[ ]	[ ]	T2b	IIB	Extension to other pelvic tissues. No malignant cells in ascites or peritoneal washings.
[ ]	[ ]	T2c	IIC	Pelvic extension (2a or 2b) with malignant cells in ascites or peritoneal washings.
[ ]	[ ]	T3 &/or N1	III	Tumor involves one or both ovaries with microscopically confirmed peritoneal metastasis outside the pelvis and/or regional lymph node metastasis
[ ]	[ ]	T3a	IIIA	Microscopic peritoneal metastasis beyond pelvis
[ ]	[ ]	T3b	IIIB	Macroscopic peritoneal metastasis beyond pelvis 2 cm or less in greatest dimension
[ ]	[ ]	T3c &/or N1	IIIC	Peritoneal metastasis beyond pelvis more than 2 cm in greatest dimension and/or regional lymph node metastasis
[ ]	[ ]	M1	IV	Distant metastasis (excludes peritoneal metastasis)

Lymph Node (N)

[ ] [ ]	NX — Regional lymph nodes cannot be assessed
[ ] [ ]	N0 — No regional lymph node metastasis
[ ] [ ]	N1 — Regional lymph node metastasis

Distant Metastasis (M)

[ ] [ ]	MX — Presence of distant metastasis cannot be assessed
[ ] [ ]	M0 — No distant metastasis
[ ] [ ]	M1 — Distant metastasis (excludes peritoneal metastasis)

Stage Grouping

Clin	Path	AJCC/UICC				FIGO*
[ ]	[ ]	IA	T1a	N0	M0	Stage IA
[ ]	[ ]	IB	T1b	N0	M0	Stage IB
[ ]	[ ]	IC	T1c	N0	M0	Stage IC
[ ]	[ ]	IIA	T2a	N0	M0	Stage IIA
[ ]	[ ]	IIB	T2b	N0	M0	Stage IIB
[ ]	[ ]	IIC	T2c	N0	M0	Stage IIC
[ ]	[ ]	IIIA	T3a	N0	M0	Stage IIIA
[ ]	[ ]	IIIB	T3b	N0	M0	Stage IIIB
[ ]	[ ]	IIIC	T3c	N0	M0	Stage IIIC
			Any T	N1	M0	
[ ]	[ ]	IV	Any T	Any N	M1	Stage IV

Staged by _____ M.D.
_____ Registrar
Date _____

* FIGO: *Federation Internationale de Gynecologie et d'Obstetrique*

Note: Liver capsular metastasis is T3/Stage III, liver parenchymal metastasis M1/Stage IV. Pleural effusion must have positive cytology for M1/Stage IV.

(continued on next page)

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] GB Borderline malignancy
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated
[ ] G3-G4 Poorly differentiated or undifferentiated

Histopathologic Type

The types currently recommended are serous tumors, mucinous tumors, endometrioid tumors, clear cell (mesonephroid) tumors, undifferentiated tumors, and unclassified tumors. Malignant tumors other than those of the common epithelial types are not to be included with the categories listed below. However, the more common ones—such as granulosa cell tumor, immature teratoma, dysgerminoma, and endodermal sinus tumor—may be collected and reported separately by institutions so desiring, particularly those with a pediatric population among their patients.

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- Serous cystadenocarcinomas

Mucinous cystomas
- Mucinous cystadenomas with proliferating activity of the epithelial cells and nuclear abnormalities, but with no infiltrative destructive growth (low potential or borderline malignancy)
- Mucinous cystadenocarcinomas

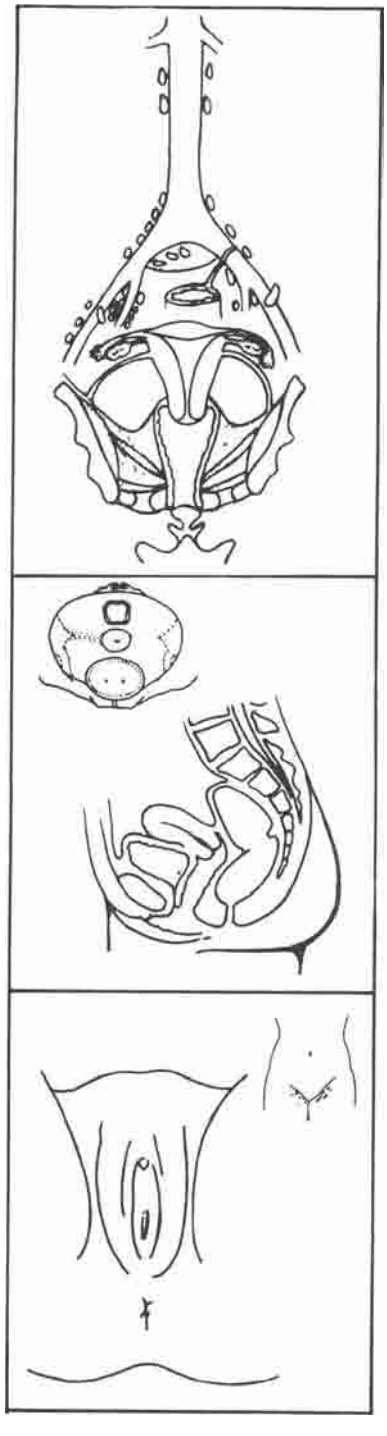
Endometrioid tumors (similar to adenocarcinomas in the endometrium)
- Endometrioid tumors with proliferating activity of the epithelial cells and nuclear abnormalities, but with no infiltrative destructive growth (low potential or borderline malignancy)
- Endometrioid adenocarcinomas

Clear cell (mesonephroid) tumors
- Clear cell tumors with proliferating activity of the epithelial cells and nuclear abnormalities, but with no infiltrative destructive growth (low potential or borderline malignancy)
- Clear cell cystadenocarcinomas

Unclassified tumors

In some cases of inoperable widespread malignant tumor, it may be impossible for the gynecologist and the pathologist to decide the origin of the growth. In order to evaluate the results obtained in the treatment of carcinoma of the ovary, however, it is necessary that all patients be reported on, including those thought to have a malignant ovarian tumor. If clinical examination cannot exclude the possibility that the lesion is a primary ovarian carcinoma, the case should be reported in the group "special category" and belong to the histologic group of unclassified tumors.

Illustrations



Indicate on diagrams primary tumor and regional nodes involved.

29

Vagina

C52.9 Vagina

ANATOMY

Primary Site. The vagina extends from the vulva upward to the uterine cervix.

Regional Lymph Nodes. They are:

Femoral (lower third only)
Inguinal (lower third only)
Common iliac
Internal iliac (hypogastric)
External iliac
Hypogastric
Pelvic, NOS (upper two-thirds only)

Metastatic Sites. The most common sites of distant spread include the lungs and skeleton.

RULES FOR CLASSIFICATION

The classification applies to primary carcinoma only.

A tumor that has extended to the portio and reached the external os should be classified as carcinoma of the cervix.

A tumor involving the vulva should be classified as carcinoma of the vulva.

There should be histologic confirmation of the disease. Any unconfirmed cases must be reported separately.

Clinical Staging. All data available prior to first definitive treatment should be used.

Pathologic Staging. In addition to data used for clinical staging, additional information available from examination of the resected specimen is to be used.

DEFINITION OF TNM

The definitions of the T categories correspond to the several stages accepted by FIGO. Both systems are included for comparison.

Primary Tumor (T)

TNM	FIGO	DEFINITION
TX	—	Primary tumor cannot be assessed
T0	—	No evidence of primary tumor
Tis	0	Carcinoma *in situ*
T1	I	Tumor confined to the vagina
T2	II	Tumor invades paravaginal tissues but not to the pelvic wall
T3	III	Tumor extends to the pelvic wall
T4*	IVA	Tumor invades the mucosa of the bladder or rectum and/or extends beyond the true pelvis
M1	IVB	Distant metastasis

**Note:* The presence of bullous edema is not sufficient evidence to classify a tumor as T4. If the mucosa is not involved, the tumor is Stage III.

Regional Lymph Nodes (N)

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

Upper Two-Thirds of the Vagina:

N1 Pelvic lymph node metastasis

Lower One-Third of the Vagina:

N1 Unilateral inguinal lymph node metastasis
N2 Bilateral inguinal lymph node metastasis

Distant Metastasis (M)

TNM	FIGO	DEFINITION
MX	—	Presence of distant metastasis cannot be assessed
M0	—	No distant metastasis
M1	IVB	Distant metastasis

pTNM Pathological Classification

The pT, pN, and pM categories correspond to the T, N, and M categories.

STAGE GROUPING

AJCC/UICC				FIGO
Stage 0	Tis	N0	M0	Stage 0
Stage I	T1	N0	M0	Stage I
Stage II	T2	N0	M0	Stage II
Stage III	T1	N1	M0	Stage III
	T2	N1	M0	
	T3	N0	M0	
	T3	N1	M0	
Stage IVA	T1	N2	M0	Stage IVA
	T2	N2	M0	
	T3	N2	M0	
	T4	Any N	M0	
Stage IVB	Any T	Any N	M1	Stage IVB

HISTOPATHOLOGIC TYPE

The squamous cell carcinoma is the most common type of cancer occurring in the vagina but infrequently an adenocarcinoma may occur in the upper one third.

HISTOPATHOLOGIC GRADE (G)

GX Grade cannot be assessed
G1 Well differentiated
G2 Moderately differentiated
G3 Poorly differentiated
G4 Undifferentiated

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	TNM category	FIGO* stage	DEFINITIONS
				Primary Tumor (T)
[ ]	[ ]	TX		Primary tumor cannot be assessed
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[ ]	[ ]	Tis	0	Carcinoma in situ
[ ]	[ ]	T1	I	Tumor confined to vagina
[ ]	[ ]	T2	II	Tumor invades paravaginal tissues but not to pelvic wall
[ ]	[ ]	T3	III	Tumor extends to pelvic wall
[ ]	[ ]	T4*	IVA	Tumor invades mucosa of bladder or rectum and/or extends beyond the true pelvis
[ ]	[ ]	M1	IVB	Distant metastasis
				Lymph Node (N)
[ ]	[ ]	NX		Regional lymph nodes cannot be assessed
[ ]	[ ]	N0		No regional lymph node metastasis
				Upper two-thirds of vagina:
[ ]	[ ]	N1		Pelvic lymph node metastasis
				Lower one-third of vagina:
[ ]	[ ]	N1		Unilateral inguinal lymph node metastasis
[ ]	[ ]	N2		Bilateral inguinal lymph node metastasis
				Distant Metastasis (M)
[ ]	[ ]	MX		Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0		No distant metastasis
[ ]	[ ]	M1	IVb	Distant metastasis

Clin	Path	Stage Grouping			
[ ]	[ ]	0	Tis	N0	M0
[ ]	[ ]	I	T1	N0	M0
[ ]	[ ]	II	T2	N0	M0
[ ]	[ ]	III	T1	N1	M0
			T2	N1	M0
			T3	N0	M0
			T3	N1	M0
[ ]	[ ]	IVA	T1	N2	M0
			T2	N2	M0
			T3	N2	M0
			T4	Any N	M0
[ ]	[ ]	IVB	Any T	Any N	M1

Staged by _____ M.D.
_____ Registrar
Date _____

** Note: The presence of bullous edema is not sufficient evidence to classify a tumor T4. If the mucosa is not involved the tumor is stage III.*

(continued on next page)

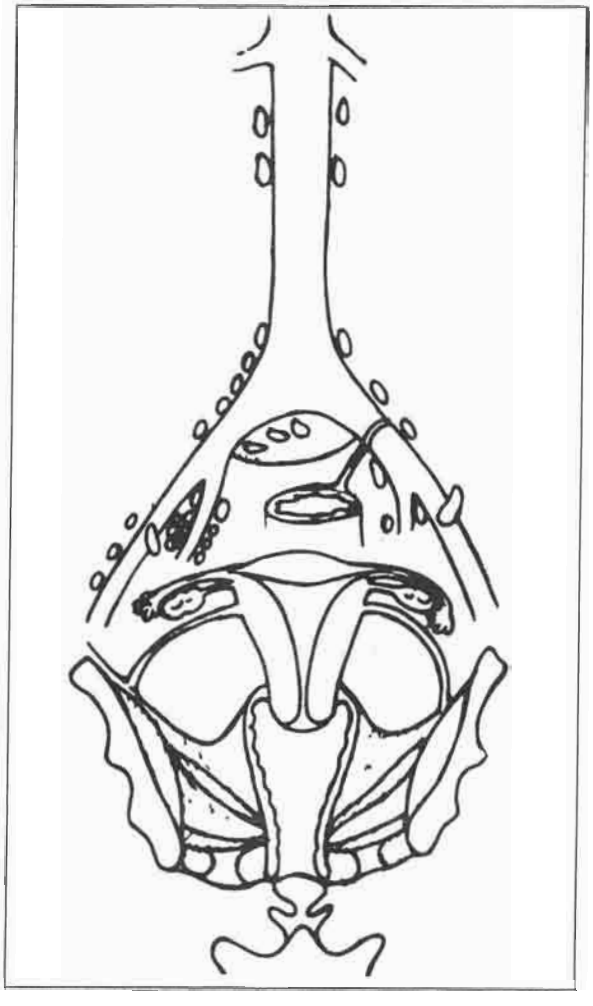
Histopathologic Type

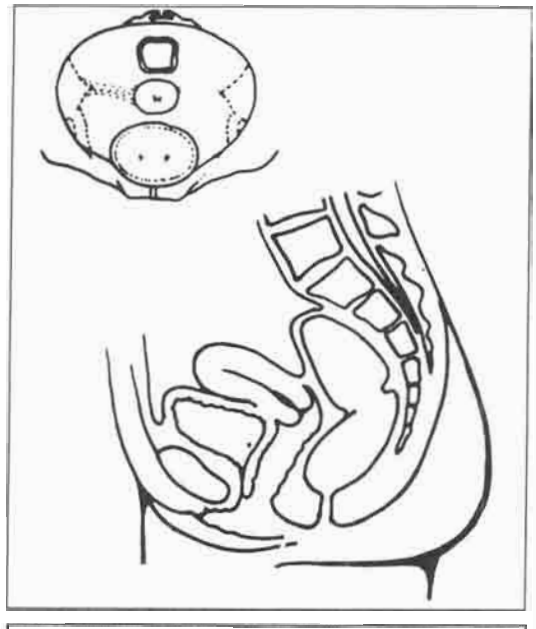
Squamous cell carcinoma
Adenocarcinoma infrequently
Upper one third

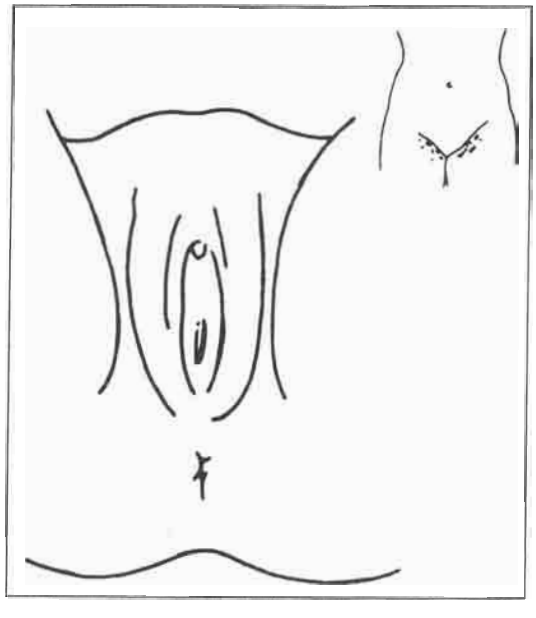
Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated
[ ] G3 Poorly differentiated
[ ] G4 Undifferentiated

Illustrations







Indicate on diagrams primary tumor and regional nodes involved.

30

Vulva

C51.0 Labium majus
C51.1 Labium minus
C51.2 Clitoris
C51.8 Overlapping lesion
C51.9 Vulva, NOS

The staging classification for carcinomas of the vulva is taken directly from FIGO.

Cases should be classified as carcinoma of the vulva when the primary site of the growth is in the vulva. Tumors present in the vulva as secondary growths from either a genital or extragenital site should be excluded. Malignant melanoma should be reported separately.

ANATOMY

Primary Site. The vulva is the anatomic area immediately external to the vagina.

Regional Lymph Nodes. The femoral and inguinal nodes are the sites of regional spread.

Metastatic Sites. These include any site beyond the area of the regional lymph nodes. Internal iliac, external iliac, and hypogastric lymph nodes are now considered distant metastasis.

RULES FOR CLASSIFICATION

The classification applies only to primary carcinoma of the vulva. There should be histologic confirmation of the cancer. A carcinoma of the vulva that has extended to the vagina should be classified as carcinoma of the vulva. Malignant melanoma should be reported separately.

Clinical Staging. The rules for staging are similar to those for carcinoma of the cervix.

Pathologic Staging. The rules of staging are similar to those for carcinoma of the cervix.

DEFINITION OF TNM

TNM classification of carcinoma of the vulva is based on the FIGO classification.

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
Tis Carcinoma *in situ* (preinvasive carcinoma)
T1 Tumor confined to the vulva or to the vulva and perineum, 2 cm or less in greatest dimension
T2 Tumor confined to the vulva or to the vulva and perineum, more than 2 cm in greatest dimension
T3 Tumor invades any of the following: lower urethra, vagina, or anus
T4 Tumor invades any of the following: bladder mucosa, upper urethral mucosa, or rectal mucosa, or is fixed to the bone

Regional Lymph Nodes (N)

NX Regional lymph nodes cannot be assessed
N0 No regional lymph node metastasis
N1 Unilateral regional lymph node metastasis
N2 Bilateral regional lymph node metastasis

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis (Pelvic lymph node metastasis is M1.)

STAGE GROUPING

(Correlation of the FIGO, UICC, and AJCC nomenclatures)

AJCC/UICC				FIGO
Stage 0	Tis	N0	M0	
Stage I	T1	N0	M0	Stage I
Stage II	T2	N0	M0	Stage II
Stage III	T1	N1	M0	Stage III
	T2	N1	M0	
	T3	N0	M0	
	T3	N1	M0	
Stage IVA	T1	N2	M0	Stage IVA
	T2	N2	M0	
	T3	N2	M0	
	T4	Any N	M0	
Stage IVB	Any T	Any N	M1	Stage IVB

HISTOPATHOLOGIC TYPE

Squamous cell carcinoma is the most common form of cancer of the vulva. Malignant melanoma should be reported separately.

The histopathologic types are:

Vulvar intraepithelial neoplasia, grade III
Squamous cell carcinoma *in situ*
Squamous cell carcinoma
Verrucous carcinoma
Paget's disease of the vulva
Adenocarcinoma, NOS
Basal cell carcinoma, NOS
Bartholin's gland carcinoma

HISTOPATHOLOGIC GRADE (G)

GX Grade cannot be assessed
G1 Well differentiated
G2 Moderately differentiated
G3 Poorly differentiated
G4 Undifferentiated

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age ____ Sex ____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	DEFINITIONS	
		Primary Tumor (T)	
[ ]	[ ]	TX	Primary tumor cannot be assessed
[ ]	[ ]	T0	No evidence of primary tumor
[ ]	[ ]	Tis	Preinvasive carcinoma (carcinoma *in situ*)
[ ]	[ ]	T1	Tumor confined to the vulva or to the vulva and perineum, 2 cm or less in greatest dimension
[ ]	[ ]	T2	Tumor confined to the vulva or to the vulva and perineum, more than 2 cm in greatest dimension
[ ]	[ ]	T3	Tumor involves any of the following: lower urethra, vagina, or anus
[ ]	[ ]	T4	Tumor invades any of the following: bladder mucosa, upper part of urethral mucosa, rectal mucosa or tumor fixed to the bone
		Lymph Node (N)	
		Regional lymph nodes are the femoral and inguinal nodes	
[ ]	[ ]	NX	Regional lymph nodes cannot be assessed
[ ]	[ ]	N0	No regional lymph node metastasis ~~bile~~
[ ]	[ ]	N1	Unilateral regional lymph node metastasis
[ ]	[ ]	N2	Bilateral regional lymph node metastasis
		Distant Metastasis (M)	
[ ]	[ ]	MX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0	No distant metastasis
[ ]	[ ]	M1	Distant metastasis (Pelvic lymph node metastasis is M1)

Clin	Path	Stage Grouping AJCC/UICC				Figo
[ ]	[ ]	0	Tis	N0	M0	
[ ]	[ ]	I	T1	N0	M0	I
[ ]	[ ]	II	T2	N0	M0	II
[ ]	[ ]	III	T1	N1	M0	III
			T2	N1	M0	
			T3	N0	M0	
			T3	N1	M0	
[ ]	[ ]	IVA	T1	N2	M0	IVA
			T2	N2	M0	
			T3	N2	M0	
			T4	Any N	M0	
[ ]	[ ]	IVB	Any T	Any N	M1	IVB

Staged by _____ M.D.
_____ Registrar
Date _____

(continued on next page)

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated
[ ] G3 Poorly differentiated
[ ] G4 Undifferentiated

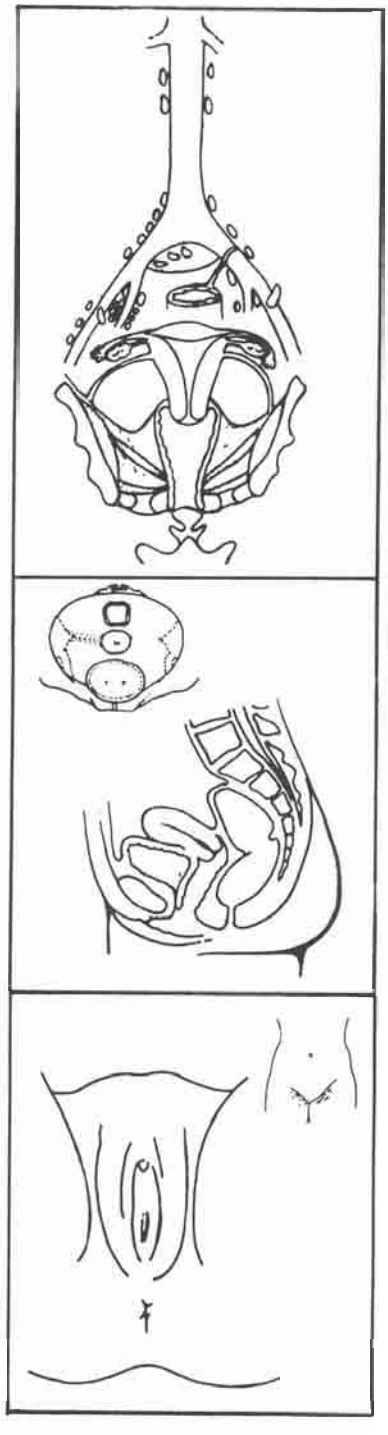
Histopathologic Type

Squamous cell carcinoma is the most frequent form of cancer of the vulva. Malignant melanoma should be reported separately.

The histopathologic types are:

Vulvar intraepithelial neoplasia, grade III
Squamous cell carcinoma *in situ*
Squamous cell carcinoma
Verrucous carcinoma
Paget's disease of the vulva
Adenocarcinoma, NOS
Basal cell carcinoma, NOS
Bartholin's gland carcinoma

Illustrations



Indicate on diagrams primary tumor and regional nodes involved.

GENITOURINARY CANCERS

31

Prostate

C61.9 Prostate gland

Prostatic cancer is the most common cancer in men, with increasing incidence in older age groups. Carcinomas of the prostate are responsive to sex hormones and presumably, therefore, have many analogies with breast cancer. They are stimulated by androgens and inhibited by estrogens. Prostatic cancer has a tendency to metastasize to bone. Early detection may now be possible with a blood test, (prostate specific antigen, PSA) and transrectal ultrasound. This TNM classification for carcinomas of the prostate is new.

ANATOMY

Primary Site. Adenocarcinomas of the prostate usually arise within the peripheral zone and are less commonly seen in the benign hyperplastic enlargement that occurs around the prostatic urethra in older men. Pathologically, cancers of the prostate are often multifocal in origin. They usually start in the peripheral posterior portion of the gland and therefore are amenable to early detection by rectal examination or by transrectal ultrasound.

There is agreement that the incidence of both clinical and latent carcinoma increases with age. However, this cancer is rarely diagnosed in men under 40 years of age. The size or extent of a localized prostatic tumor may be estimated by digital examination or by various imaging techniques, such as ultrasound. Diagnosis of clinically suspicious areas of the prostate is histologically confirmed by needle biopsy.

The grade of the prostatic cancer is also important for prognosis. The histopathologic grading of these tumors can be complex because of the morphologic heterogeneity so often encountered in surgical specimens. Either a histologic or a pattern type of grading method can be used.

Regional Lymph Nodes. The regional lymph nodes are the nodes of the true pelvis, which essentially are the pelvic nodes below the bifurcation of the common iliac arteries. They include the following groups:

Pelvic, NOS
Hypogastric
Obturator
Iliac (internal, external, NOS)
Periprostatic
Sacral (lateral, presacral, promontory [Gerota's], or NOS)

Distant Lymph Nodes. Distant lymph nodes lie outside the confines of the true pelvis. They can be imaged using ultrasound, computed tomography, magnetic resonance imaging, or lymphangiography.

Aortic (para-aortic, lumbar)
Common iliac
Inguinal, deep
Superficial inguinal (femoral)
Supraclavicular
Cervical
Scalene
Retroperitoneal, NOS

The significance of regional lymph node metastasis, pN, in staging prostate cancer lies in the number of nodes involved with tumor and the size of the metastatic foci present within the lymph nodes.

Metastatic Sites. Metastasis to bone is common with primary carcinomas of the prostate. In addition, tumor frequently spreads to distant lymph nodes. Lung metastases are uncommon and may be lymphangitic in pattern of spread. Liver metastases are usually seen late in the course of the disease.

RULES FOR CLASSIFICATION

The TNM classification serves both clinical and pathological staging.

Clinical Staging. Primary tumor assessment includes digital rectal examination of the prostate and histologic or cytologic confirmation of prostatic carcinoma. Clinical examination, acid phosphatase determination, PSA serum level, and imaging techniques (including transrectal ultrasound) are suggested. All information available prior to first definitive treatment may be used for clinical staging.

Pathologic Staging. Histologic examination of the resected specimen is required. Total prostatoseminalvesiculectomy and pelvic lymph node dissection are required for pathologic staging. In some cases, a pT classification may be possible without prostatoseminalvesiculectomy—for example, a positive biopsy from the rectum. Tumor found in one or both lobes by needle biopsy, but not palpable or visible by imaging, is classified as T1c. Laterality does not affect the N classification.

DEFINITION OF TNM

Primary Tumor (T)

- TX Primary tumor cannot be assessed
- T0 No evidence of primary tumor
- T1 Clinically inapparent tumor not palpable or visible by imaging
  - T1a Tumor incidental histologic finding in 5% or less of tissue resected
  - T1b Tumor incidental histologic finding in more than 5% of tissue resected
  - T1c Tumor identified by needle biopsy (e.g., because of elevated PSA)
- T2 Tumor confined within the prostate*
  - T2a Tumor involves half of a lobe or less
  - T2b Tumor involves more than half of a lobe, but not both lobes
  - T2c Tumor involves both lobes
- T3 Tumor extends through the prostatic capsule**
  - T3a Unilateral extracapsular extension
  - T3b Bilateral extracapsular extension
  - T3c Tumor invades the seminal vesicle(s)
- T4 Tumor is fixed or invades adjacent structures other than the seminal vesicles
  - T4a Tumor invades any of: bladder neck, external sphincter, or rectum
  - T4b Tumor invades levator muscles and/or is fixed to the pelvic wall

**Note:* Tumor found in one or both lobes by needle biopsy, but not palpable or visible by imaging, is classified as T1c.

***Note:* Invasion into the prostatic apex or into (but not beyond) the prostatic capsule is not classified as T3, but as T2.

Regional Lymph Nodes (N)

- NX Regional lymph nodes cannot be assessed
- N0 No regional lymph node metastasis
- N1 Metastasis in a single lymph node, 2 cm or less in greatest dimension
- N2 Metastasis in a single lymph node, more than 2 cm but not more than 5 cm in greatest dimension; or multiple lymph node metastases, none more than 5 cm in greatest dimension
- N3 Metastasis in a lymph node more than 5 cm in greatest dimension

Distant Metastasis* (M)

- MX Presence of distant metastasis cannot be assessed
- M0 No distant metastasis
- M1 Distant metastasis
  - M1a Nonregional lymph node(s)
  - M1b Bone(s)
  - M1c Other site(s)

**Note:* When more than one site of metastasis is present, the most advanced category (pM1c) is used.

STAGE GROUPING

Stage 0	T1a	N0	M0	G1
Stage I	T1a	N0	M0	G2, 3-4
	T1b	N0	M0	Any G
	T1c	N0	M0	Any G
	T1	N0	M0	Any G
Stage II	T2	N0	M0	Any G
Stage III	T3	N0	M0	Any G
Stage IV	T4	N0	M0	Any G
	Any T	N1	M0	Any G
	Any T	N2	M0	Any G
	Any T	N3	M0	Any G
	Any T	Any N	M1	Any G

HISTOPATHOLOGIC TYPE

This classification applies to adenocarcinoma, but not to sarcoma or transitional cell carcinoma of the prostate.

HISTOPATHOLOGIC GRADE (G)

- GX Grade cannot be assessed
- G1 Well differentiated (slight anaplasia)
- G2 Moderately differentiated (moderate anaplasia)
- G3-4 Poorly differentiated or undifferentiated (marked anaplasia)

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Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	DEFINITIONS
		Primary Tumor (T)
[ ]	[ ]	TX Primary tumor cannot be assessed
[ ]	[ ]	T0 No evidence of primary tumor
[ ]	[ ]	T1 Clinically inapparent tumor not palpable or visible by imaging
[ ]	[ ]	T1a Tumor incidental histologic finding in 5% or less of tissue resected
[ ]	[ ]	T1b Tumor incidental histologic finding in more than 5% of tissue resected
[ ]	[ ]	T1c Tumor identified by needle biopsy (*e.g.* because of elevated PSA)
[ ]	[ ]	T2 Palpable tumor confined within prostate*
[ ]	[ ]	T2a Tumor involves half of a lobe or less
[ ]	[ ]	T2b Tumor involves more than half of a lobe, but not both lobes
[ ]	[ ]	T2c Tumor involves both lobes
[ ]	[ ]	T3 Tumor extends through the prostatic capsule**
[ ]	[ ]	T3a Unilateral extracapsular extension
[ ]	[ ]	T3b Bilateral extracapsular extension
[ ]	[ ]	T3c Tumor invades seminal vesicle(s)
[ ]	[ ]	T4 Tumor is fixed or invades adjacent structures other than seminal vesicles
[ ]	[ ]	T4a Tumor invades external sphincter and/or bladder neck and/or rectum
[ ]	[ ]	T4b Tumor invades levator muscles and/or is fixed to pelvic wall
		Lymph Node (N)
[ ]	[ ]	NX Regional lymph nodes cannot be assessed
[ ]	[ ]	N0 No regional lymph node metastasis
[ ]	[ ]	N1 Metastasis in a single lymph node, 2 cm or less in greatest dimension
[ ]	[ ]	N2 Metastasis in a single lymph node, more than 2 cm but not more than 5 cm in greatest dimension, or multiple lymph nodes, none more than 5 cm in greatest dimension
[ ]	[ ]	N3 Metastasis in a lymph node more than 5 cm in greatest dimension
		Distant Metastasis (M)* **
[ ]	[ ]	MX Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0 No distant metastasis
[ ]	[ ]	M1 Distant metastasis
[ ]	[ ]	M1a Non-regional lymph nodes
[ ]	[ ]	M1b Bone
[ ]	[ ]	M1c Other sites

* *Note:* Tumor found in one or both lobes by needle biopsy, but not palpable or visible by imaging, is classified as T1c.
** *Note:* Invasion into the prostatic apex or into (but not beyond) the prostatic capsule is not classified as T3, but as T2.
*** *Note:* When more than one site of metastasis is present, the most advanced category (pM1c) is used.

(continued on next page)

Clin	Path	Stage Grouping				
[ ]	[ ]	0	T1a	N0	M0	G1
[ ]	[ ]	I	T1a	N0	M0	G2
			T1a	N0	M0	G3-4
			T1b	N0	M0	Any G
			T1c	N0	M0	Any G
[ ]	[ ]		T1	N0	M0	Any G
[ ]	[ ]	II	T2	N0	M0	Any G
[ ]	[ ]	III	T3	N0	M0	Any G
		IV	T4	N0	M0	Any G
			Any T	N1	M0	Any G
			Any T	N2	M0	Any G
			Any T	N3	M0	Any G
			Any T	Any N	M1	Any G

Staged by _____ M.D.

_____ Registrar

Date _____

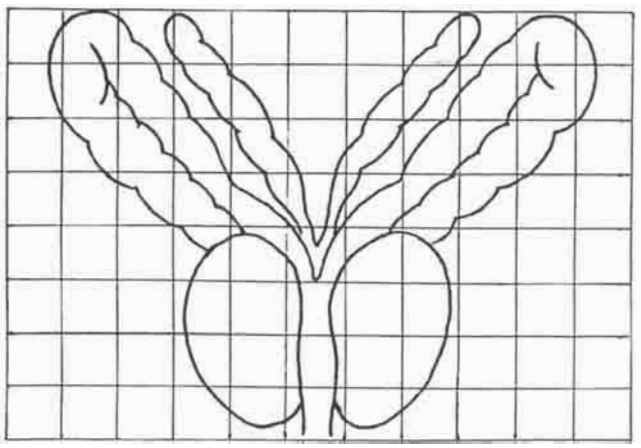
Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated (slight anaplasia)
[ ] G2 Moderately well differentiated (moderate anaplasia)
[ ] G3-4 Poorly differentiated or undifferentiated (marked anaplasia)

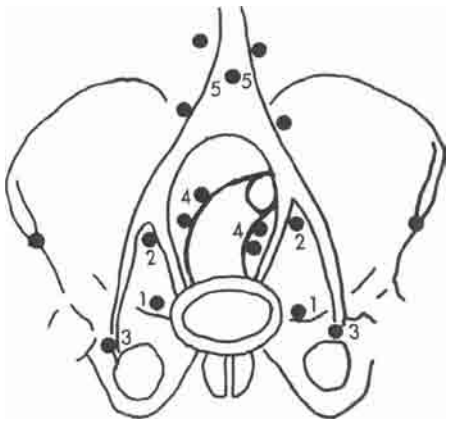
Histopathologic Type

Almost always adenocarcinoma of variable grades.

Illustrations



This diagram is for use with the prostate diagram. Sketch in extent of tumor.



Indicate on diagram primary tumor and regional nodes involved.

32

Testis

C62.0 Undescended testis
C62.1 Descended testis
C62.9 Testis, NOS

Cancers of the testis are usually found in young adults. Fortunately, they are relatively rare, accounting for less than 1% of all malignancies in males. Cryptorchidism is a predisposing condition. There are two main histologic types: seminomas, which are most common, and teratomas. Most cases of testicular cancer, even when far advanced, can be successfully treated. Circulating tumor markers are found in the serum of patients with cancer of the testis, which enables the clinician to document the course of the disease. These markers are invaluable for the management of testicular malignancies. Staging is based on the extent of disease.

ANATOMY

Primary Site. The testes are composed of convoluted seminiferous tubules with a stroma containing functional endocrine interstitial cells. Both are encased in a dense barrier capsule, the tunica albuginea, with fibrous septa extending into and separating the testes into lobules. The tubules converge and exit at the mediastinum of the testis into the rete testis and efferent ducts, which join a single duct. This duct—the epididymis—coils outside the upper and lower pole of the testicle, then joins the vas deferens, a muscular conduit that accompanies the vessels and lymphatic channels of the spermatic cord. The major route for local extension of cancer is through the lymphatic channels. The tumor emerges from the mediastinum of the testis and courses through the spermatic cord. Occasionally, the epididymis is invaded early, and then the external iliac nodes may become involved. If there has been previous scrotal or inguinal surgery with invasion of the scrotal wall (though this is rare), then the lymphatic spread may be to inguinal nodes.

Regional Lymph Nodes. The regional lymph nodes are:

Aortic
Paraaortic
External ileac
Paracaval
Intrapelvic
Inguinal (after scrotal or inguinal surgery)

Spread of the tumor into contralateral regional or first station nodes of the area occurs in 20% of cases. When there has been previous inguinal or scrotal surgery, inguinal nodes are also considered regional nodes. All nodes outside the regional nodes are distant. As defined, bulky disease has important prognostic significance.

The significance of regional lymph node metastasis in staging testicular cancer lies in the number and size and not in whether metastasis is unilateral or contralateral.

Metastatic Sites. Distant spread of testicular tumors occurs most commonly to the nodes, followed by metastasis to the lung, liver, viscera, and bones. As defined, bulk of disease has important prognostic significance. Serum markers (alphafetoprotein [AFP]) and the beta-subunit of human chorionic gonadotropin (βHCG) should be obtained prior to initial orchiectomy to establish whether the tumor marker is predictive. Markers are helpful in the management of patients with disseminated disease. Stage can be further subdivided by the presence or absence of markers.

RULES FOR CLASSIFICATION

Clinical Staging. Clinical examination and radical orchiectomy are required for clinical staging.

Pathologic Staging. Histologic evaluation of the radical orchiectomy specimen must be used for the pT clas-

sification. The specimens from a defined node-bearing area (e.g., retroperitoneal periaortic node dissection) must be used for the pN classification. Histologic verification is required. Laterality does not affect the N classification.

DEFINITION OF TNM

Primary Tumor (T)

The extent of primary tumor is classified after radical orchiectomy.

- pTX Primary tumor cannot be assessed (If no radical orchiectomy has been performed, TX is used.)
- pT0 No evidence of primary tumor (e.g., histologic scar in testis)
- pTis Intratubular tumor: preinvasive cancer
- pT1 Tumor limited to the testis, including the rete testis
- pT2 Tumor invades beyond the tunica albuginea or into the epididymis
- pT3 Tumor invades the spermatic cord
- pT4 Tumor invades the scrotum

Regional Lymph Nodes (N)

- NX Regional lymph nodes cannot be assessed
- N0 No regional lymph node metastasis
- N1 Metastasis in a single lymph node, 2 cm or less in greatest dimension
- N2 Metastasis in a single lymph node, more than 2 cm but not more than 5 cm in greatest dimension; or multiple lymph nodes, none more than 5 cm in greatest dimension
- N3 Metastasis in a lymph node more than 5 cm in greatest dimension

Distant Metastasis (M)

- MX Presence of distant metastasis cannot be assessed
- M0 No distant metastasis
- M1 Distant metastasis

STAGE GROUPING

Stage 0	pTis	N0	M0
Stage I	Any pT	N0	M0
Stage II	Any pT	N1	M0
	Any pT	N2	M0
	Any pT	N3	M0
Stage III	Any pT	Any N	M1

HISTOPATHOLOGIC TYPE

Cell types can be divided into seminomatous and nonseminomatous tumors. The latter can be further divided into teratoma, embryonal carcinoma, yolk sac tumor, and choriocarcinoma. Mixtures of these types should be noted. Lymphomas are excluded. Combinations of embryonal carcinoma and teratoma can be designated as teratocarcinoma.

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Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	DEFINITIONS
		Primary Tumor (T)
[ ]	[ ]	pTX Primary tumor cannot be assessed. (If no radical orchiectomy has been performed, TX is used)
[ ]	[ ]	pT0 No evidence of primary tumor (*e.g.* histologic scar in testis)
[ ]	[ ]	pTis Intratubular tumor: preinvasive cancer
[ ]	[ ]	pT1 Tumor limited to testis, including rete testis
[ ]	[ ]	pT2 Tumor invades beyond tunica albuginea or into epididymis
[ ]	[ ]	pT3 Tumor invades spermatic cord
[ ]	[ ]	pT4 Tumor invades scrotum
		Lymph Node (N)
[ ]	[ ]	NX Regional lymph nodes cannot be assessed
[ ]	[ ]	N0 No regional lymph node metastasis
[ ]	[ ]	N1 Metastasis in a single lymph node, 2 cm or less in greatest dimension
[ ]	[ ]	N2 Metastasis in a single lymph node, more than 2 cm but not more than 5 cm in greatest dimension, or multiple lymph nodes, none more than 5 cm in greatest dimension
[ ]	[ ]	N3 Metastasis in a lymph node more than 5 cm in greatest dimension
		Distant Metastasis (M)
[ ]	[ ]	MX Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0 No distant metastasis
[ ]	[ ]	M1 Distant metastasis

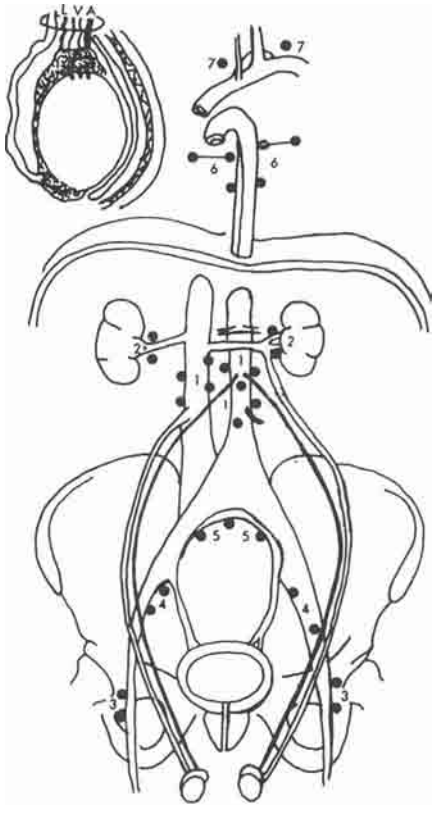
Clin	Path	Stage Grouping			
]	[ ]	0	pTis	N0	M0
]	[ ]	I	Any pT	N0	M0
[ ]	[ ]	II	Any pT	N1	M0
			Any pT	N2	M0
			Any pT	N3	M0
[ ]	[ ]	III	Any pT	Any N	M1

Staged by _____ M.D.
_____ Registrar
Date _____

Histopathologic Type

Cell types can be divided into seminomatous and nonseminomatous tumors. The latter can be further divided into teratoma, embryonal carcinoma, yolk sac tumor, and choriocarcinoma. Mixtures of these types should be noted. Lymphomas are excluded. Combinations of embryonal carcinoma and teratoma can be designated as teratocarcinoma.

Illustrations



Indicate on diagram primary tumor and regional nodes involved.

33

Penis

C60.0 Prepuce
C60.1 Glans penis
C60.2 Body of penis
C60.8 Overlapping lesion
C60.9 Penis, NOS

Cancers of the penis are rare in the United States, and incidence varies in different countries of the world. Most are squamous cell carcinomas arising in the skin or on the glans penis. Prognosis is favorable, provided the lymph nodes are not involved. Melanomas can also occur. The staging classification, however, applies to carcinomas. (Melanomas are staged in Chapter 24.) Some cancers of the penis may be described as verrucous. These are included under this classification. An *in situ* lesion is also included and by definition should be coded as an *in situ* carcinoma of the penis.

ANATOMY

Primary Site. The penis is composed of three cylindrical masses of cavernous tissue bound by fibrous tissue. Two masses are lateral, known as the corpora cavernosa penis. The corpus spongiosum penis, a median mass, contains the greater part of the urethra. The penis is attached to the front and the sides of the pubic arch. The skin covering the penis is thin and loosely connected with the deeper parts of the organ. This skin at the root of the penis is continuous with that over the scrotum and perineum. Distally, the skin folds upon itself to form the prepuce or foreskin. Circumcision has been associated with decreased incidence of cancer of the penis.

Regional Lymph Nodes. The regional lymph nodes are:

Single superficial inguinal (femoral)
Multiple or bilateral superficial inguinal (femoral)
Deep inguinal: Rosenmuller's or Cloquet's node
External iliac
Internal iliac (hypogastric)
Pelvic nodes, NOS

Metastatic Sites. Lung, liver, or bone are most often involved.

RULES FOR CLASSIFICATION

Clinical Staging. Clinical examination, endoscopy (where possible), and histologic confirmation are required. Imaging techniques are indicated for metastatic disease detection.

Pathologic Staging. Complete resection of the primary site with appropriate margins is required. Where regional lymph node involvement is suspected, these should be included.

DEFINITION OF TNM

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
Tis Carcinoma *in situ*
Ta Noninvasive verrucous carcinoma
T1 Tumor invades subepithelial connective tissue
T2 Tumor invades the corpus spongiosum or cavernosum
T3 Tumor invades the urethra or prostate
T4 Tumor invades other adjacent structures

Regional Lymph Nodes (N)

NX Regional lymph nodes cannot be assessed
N0 No regional lymph node metastasis
N1 Metastasis in a single superficial inguinal lymph node

N2 Metastasis in multiple or bilateral superficial inguinal lymph nodes
N3 Metastasis in deep inguinal or pelvic lymph node(s), unilateral or bilateral

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

STAGE GROUPING

Stage 0	Tis	N0	M0
	Ta	N0	M0
Stage I	T1	N0	M0
Stage II	T1	N1	M0
	T2	N0	M0
	T2	N1	M0
Stage III	T1	N2	M0
	T2	N2	M0
	T3	N0	M0
	T3	N1	M0
	T3	N2	M0
Stage IV	T4	Any N	M0
	Any T	N3	M0
	Any T	Any N	M1

HISTOPATHOLOGIC TYPE

Cell types are limited to carcinomas.

HISTOPATHOLOGIC GRADE (G)

GX Grade cannot be assessed
G1 Well differentiated
G2 Moderately differentiated
G3-4 Poorly differentiated or Undifferentiated

BIBLIOGRAPHY

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Data Form for Cancer Staging

Patient identification
Name ______________________
Address ______________________
Hospital or clinic number ______________________
Age _____ Sex _____ Race ______________________

Institution identification
Hospital or clinic ______________________
Address ______________________

Oncology Record

Anatomic site of cancer ______________________
Histologic type ______________________
Grade (G) ______________________
Date of classification ______________________

Chronology of classification
[ ] **Clinical (use all data** prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

DEFINITIONS

Primary Tumor (T)

Clin	Path		
[ ]	[ ]	TX	Primary tumor cannot be assessed
[ ]	[ ]	T0	No evidence of primary tumor
[ ]	[ ]	Tis	Carcinoma *in situ*
[ ]	[ ]	Ta	Noninvasive verrucous carcinoma
[ ]	[ ]	T1	Tumor invades subepithelial connective tissue
[ ]	[ ]	T2	Tumor invades corpus spongiosum or cavernosum
[ ]	[ ]	T3	Tumor invades urethra or prostate
[ ]	[ ]	T4	Tumor invades other adjacent structures

Lymph Node (N)

Clin	Path		
[ ]	[ ]	NX	Regional lymph nodes cannot be assessed
[ ]	[ ]	N0	No regional lymph node metastasis
[ ]	[ ]	N1	Metastasis in a single, superficial inguinal lymph node
[ ]	[ ]	N2	Metastasis in multiple or bilateral superficial inguinal lymph nodes
[ ]	[ ]	N3	Metastasis in deep inguinal or pelvic lymph node(s), unilateral or bilateral

Distant Metastasis

Clin	Path		
[ ]	[ ]	MX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0	No distant metastasis
[ ]	[ ]	M1	Distant metastasis

Stage Grouping

Clin	Path				
[ ]	[ ]	0	Tis	N0	M0
			Ta	N0	M0
[ ]	[ ]	I	T1	N0	M0
[ ]	[ ]	II	T1	N1	M0
			T2	N0	M0
			T2	N1	M0
[ ]	[ ]	III	T1	N2	M0
			T2	N2	M0
			T3	N0	M0
			T3	N1	M0
			T3	N2	M0
[ ]	[ ]	IV	T4	Any N	M0
			Any T	N3	M0
			Any T	Any N	M1

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated
[ ] G3-4 Poorly differentiated or undifferentiated

Histopathologic Type

Cell types are limited to carcinomas.

Staged by ______________________ M.D.
______________________ Registrar
Date ______________________

34

Urinary Bladder

C67.0 Trigone
C67.1 Dome
C67.2 Lateral wall
C67.3 Anterior wall
C67.4 Posterior wall
C67.5 Bladder neck
C67.6 Ureteric orifice
C67.7 Urachus
C67.8 Overlapping lesion
C67.9 Bladder, NOS

Bladder cancer can present as a low grade papillary lesion, as an indolent *in situ* lesion, which can occupy large areas of the mucosal surface, or as an infiltrative cancer that rapidly extends through the bladder wall. The papillary and *in situ* lesions may be associated with a malignant course, with sudden invasion of the bladder wall. Predisposing factors include exposure to certain chemicals used in the dye industry, and smoking. Bladder cancer is more common in men. Hematuria is the most common presenting sign.

ANATOMY

Primary Site. The urinary bladder consists of three layers: the epithelium and the subepithelial connective tissue, the muscularis, and the perivesical fat (peritoneum covering the superior surface and upper part). In the male, the bladder adjoins the rectum and seminal vesicle posteriorly, the prostate inferiorly, and the pubis and peritoneum anteriorly. In the female, the vagina is located posteriorly and the uterus superiorly. The bladder is located extraperitoneally.

Regional Lymph Nodes. The regional lymph nodes are the nodes of the true pelvis, which essentially are the pelvic nodes below the bifurcation of the common iliac arteries.

The significance of regional lymph node metastasis in staging bladder cancer lies in the number and size and not in whether metastasis is unilateral or contralateral.

Regional nodes include:

Hypogastric
Obturator
Iliac (internal, external, NOS)
Perivesical
Pelvic, NOS
Sacral (lateral, sacral promontory [Gerota's])
Presacral

The common iliac nodes are considered sites of distant metastasis and should be coded as M1.

Metastatic Sites. Distant spread to lymph nodes, lung, bone, and liver is most common.

RULES FOR CLASSIFICATION

Clinical Staging. Primary tumor assessment includes bimanual examination under anesthesia before and after endoscopic surgery (biopsy or transurethral resection) or histologic verification of the presence or absence of tumor when indicated. Add "m" for multiple tumors. Add "is" to any T to indicate associated carcinoma *in situ*. Appropriate imaging techniques for lymph node evaluation should be used. When indicated, evaluation for distant metastases includes imaging of the chest, biochemical studies, and isotopic studies to detect common metastatic sites. Computed tomography or other modalities may subsequently be used to supply information concerning minimal require-

ments for staging. The primary tumor biopsy may be superficial or invasive and can be partially or totally resected with sufficient tissue from the tumor base for full evaluation of depth of tumor invasion. Visually adjacent cystoscopically normal mucosa should be considered for biopsy; urinary cytology and pyelography are important.

Pathologic Staging. Microscopic examination and confirmation of extent is required. Total cystectomy and lymph node dissection generally are required for this staging. Laterality does not affect the N classification.

DEFINITION OF TNM

Primary Tumor (T)

- TX Primary tumor cannot be assessed
- T0 No evidence of primary tumor
- Ta Noninvasive papillary carcinoma
- Tis Carcinoma *in situ*: "flat tumor"
- T1 Tumor invades subepithelial connective tissue
- T2 Tumor invades superficial muscle (inner half)
- T3 Tumor invades deep muscle or perivesical fat
  - T3a Tumor invades deep muscle (outer half)
  - T3b Tumor invades perivesical fat
    - i. microscopically
    - ii. macroscopically (extravesical mass)
- T4 Tumor invades any of the following: prostate, uterus, vagina, pelvic wall, or abdominal wall
  - T4a Tumor invades the prostate, uterus, or vagina
  - T4b Tumor invades the pelvic wall or abdominal wall

Regional Lymph Nodes (N)

Regional lymph nodes are those within the true pelvis; all others are distant nodes.

- NX Regional lymph nodes cannot be assessed
- N0 No regional lymph node metastasis
- N1 Metastasis in a single lymph node, 2 cm or less in greatest dimension
- N2 Metastasis in a single lymph node, more than 2 cm but not more than 5 cm in greatest dimension; or multiple lymph nodes, none more than 5 cm in greatest dimension
- N3 Metastasis in a lymph node more than 5 cm in greatest dimension

Distant Metastasis (M)

- MX Presence of distant metastasis cannot be assessed
- M0 No distant metastasis
- M1 Distant metastasis

STAGE GROUPING

Stage 0a	Ta	N0	M0
Stage 0is	Tis	N0	M0
Stage I	T1	N0	M0
Stage II	T2	N0	M0
	T3a	N0	M0
Stage III	T3b	N0	M0
	T4a	N0	M0
Stage IV	T4b	N0	M0
	Any T	N1	M0
	Any T	N2	M0
	Any T	N3	M0
	Any T	Any N	M1

HISTOPATHOLOGIC TYPE

The histologic types are:

- Transitional cell carcinoma (urothelial)
  - *In situ*
    - Papillary
    - Flat
  - With squamous metaplasia
  - With glandular metaplasia
  - With squamous and glandular metaplasia
- Squamous cell carcinoma
- Adenocarcinoma
- Undifferentiated carcinoma

The predominant cancer is a transitional cell cancer.

HISTOPATHOLOGIC GRADE (G)

- GX Grade cannot be assessed
- G1 Well differentiated
- G2 Moderately differentiated
- G3-4 Poorly differentiated or undifferentiated

BIBLIOGRAPHY

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6. Smith JA Jr, Whitmore WF Jr: Regional lymph node metastasis from bladder cancer. J Urol 126:591–593, 1981
7. Whitmore WF Jr: Management of bladder cancer. Curr Probl Cancer 4:3–48, 1979

Data Form for Cancer Staging

Patient identification
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Hospital or clinic number ______________________
Age _____ Sex _____ Race ______________________

Institution identification
Hospital or clinic ______________________
Address ______________________

Oncology Record

Anatomic site of cancer ______________________
Histologic type ______________________
Grade (G) ______________________
Date of classification ______________________

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

DEFINITIONS

Primary Tumor (T)

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[ ]	[ ]	Tis	Carcinoma *in situ:* "flat tumor"
[ ]	[ ]	T1	Tumor invades subepithelial connective tissue
[ ]	[ ]	T2	Tumor invades superficial muscle (inner half)
[ ]	[ ]	T3	Tumor invades deep muscle or perivesical fat
[ ]	[ ]	T3a	Tumor invades deep muscle (outer half)
[ ]	[ ]	T3b	Tumor invades perivesical fat i. microscopically ii. macroscopically (extravesical mass)
[ ]	[ ]	T4	Tumor invades prostate, uterus, vagina, pelvic wall or abdominal wall
[ ]	[ ]	T4a	Tumor invades prostate, uterus, vagina
[ ]	[ ]	T4b	Tumor invades pelvic wall or abdominal wall

Lymph Node (N)

Clin	Path		
[ ]	[ ]	NX	Regional lymph nodes cannot be assessed
[ ]	[ ]	N0	No regional lymph node metastasis
[ ]	[ ]	N1	Metastasis in a single lymph node, 2 cm or less in greatest dimension
[ ]	[ ]	N2	Metastasis in a single lymph node, more than 2 cm but not more than 5 cm in greatest dimension, or multiple lymph nodes, none more than 5 cm in greatest dimension
[ ]	[ ]	N3	Metastasis in a lymph node more than 5 cm in greatest dimension

Distant Metastasis (M)

Clin	Path		
[ ]	[ ]	MX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0	No distant metastasis
[ ]	[ ]	M1	Distant metastasis

Stage Grouping

Clin	Path				
[ ]	[ ]	0a	Ta	N0	M0
[ ]	[ ]	0is	Tis	N0	M0
[ ]	[ ]	I	T1	N0	M0
[ ]	[ ]	II	T2	N0	M0
			T3a	N0	M0
[ ]	[ ]	III	T3b	N0	M0
			T4a	N0	M0
[ ]	[ ]	IV	T4b	N0	M0
			Any T	N1	M0
			Any T	N2	M0
			Any T	N3	M0
			Any T	Any N	M1

Staged by ______________________ M.D.
______________________ Registrar
Date ______________________

(continued on next page)

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated
[ ] G3-4 Poorly differentiated or undifferentiated

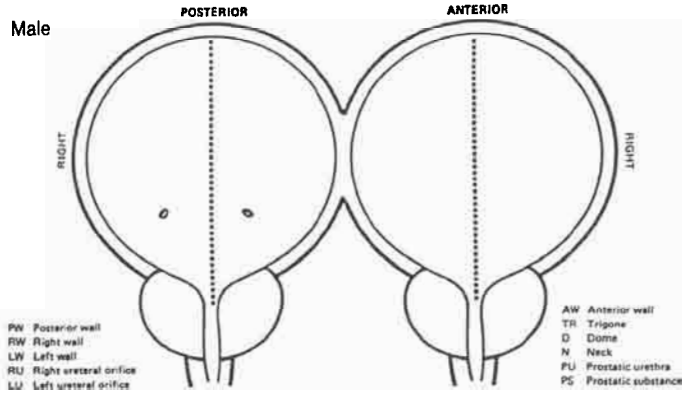
Histopathologic Type

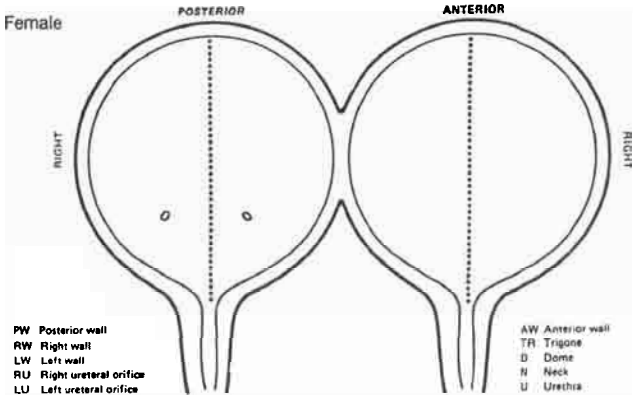
The histologic types are:

Transitional cell carcinoma (urothelial)
In situ
Papillary
Flat
With squamous metaplasia
With glandular metaplasia
With squamous and glandular metaplasia
Squamous cell carcinoma
Adenocarcinoma
Undifferentiated carcinoma

The predominant cancer is a transitional cell cancer.

Illustrations





Indicate on diagrams primary tumor and regional nodes involved.

35

Kidney

C64.9 Kidney, NOS

Cancers of the kidney are relatively rare, accounting for less than 3% of all malignancies. Nearly all malignant tumors are carcinomas arising from the renal tubular epithelium or, less frequently, from the renal pelvis (see chapter 36). These tumors are more common in males. Pain and hematuria are the common presenting features. Renal carcinomas may be associated with erythrocytosis, secondary to release of erythropoietin from the tumor cells. These carcinomas have a tendency to extend along the renal vein and even into the vena cava. Rarely, they also may regress spontaneously. Staging depends on the size of the primary tumor, invasion of the adjacent structures, and vascular extension.

ANATOMY

Primary Site. Encased by a fibrous capsule and surrounded by perirenal fat, the kidney consists of the cortex (glomeruli, convoluted tubules) and the medulla (Henle's loops, pyramids of converging tubules). Each papilla opens into the minor calices; these in turn unite in the major calices and drain into the renal pelvis. At the hilus are the pelvis, ureter, and renal artery and vein. Gerota's fascia overlies the psoas and quadrants lumborum.

Regional Lymph Nodes. The regional lymph nodes include:

Renal hilar
Paracaval
Aortic (para-aortic, periaortic, lateral aortic)
Retroperitoneal, NOS

Metastatic Sites. Common metastatic sites include bone, liver, lung, brain, and distant nodes.

RULES FOR CLASSIFICATION

The classification applies only to the renal-cell carcinomas. Adenoma is excluded. There should be histologic confirmation of the disease. Refer to Histopathologic Type.

Clinical Staging. Clinical examination, urography, and appropriate imaging techniques are required for assessment of the primary tumor and its extensions, both local and distant. Evaluation for distant metastases should be done by laboratory biochemical studies, chest x-rays, and isotopic studies. Clinical staging may also include laparotomy and biopsy of distant sites.

Pathologic Staging. Histologic examination and confirmation of extent is required. Resection of the primary tumor, kidney, Gerota's fascia, perinephric fat, renal vein, and appropriate lymph nodes is required. Laterality does not affect the N classification.

DEFINITION OF TNM

Primary Tumor (T)

- TX Primary tumor cannot be assessed
- T0 No evidence of primary tumor
- T1 Tumor 2.5 cm or less in greatest dimension limited to the kidney
- T2 Tumor more than 2.5 cm in greatest dimension limited to the kidney
- T3 Tumor extends into major veins or invades the adrenal gland or perinephric tissues but not beyond Gerota's fascia
  - T3a Tumor invades the adrenal gland or perinephric tissues but not beyond Gerota's fascia
  - T3b Tumor grossly extends into the renal vein(s) or vena cava below the diaphragm
  - T3c Tumor grossly extends into the vena cava above the diaphragm
- T4 Tumor invades beyond Gerota's fascia

Regional Lymph Nodes (N)*

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

N1 Metastasis in a single lymph node, 2 cm or less in greatest dimension
N2 Metastasis in a single lymph node, more than 2 cm but not more than 5 cm in greatest dimension; or multiple lymph nodes, none more than 5 cm in greatest dimension
N3 Metastasis in a lymph node more than 5 cm in greatest dimension

* *Note:* Laterality does not affect the N classification.

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

STAGE GROUPING

Stage I	T1	N0	M0
Stage II	T2	N0	M0
Stage III	T1	N1	M0
	T2	N1	M0
	T3a	N0	M0
	T3a	N1	M0
	T3b	N0	M0
	T3b	N1	M0
	T3c	N0	M0
	T3c	N1	M0
Stage IV	T4	Any N	M0
	Any T	N2	M0
	Any T	N3	M0
	Any T	Any N	M1

HISTOPATHOLOGIC TYPE

The histopathologic types are:

Renal cell carcinoma
Adenocarcinoma
Renal papillary adenocarcinoma
Tubular carcinoma
Granular cell carcinoma
Clear cell carcinoma (hypernephroma)

The predominant cancer is adenocarcinoma; subtypes are clear-cell and granular-cell carcinoma. A grading system as provided below is recommended when feasible. The staging system does not apply to sarcomas of the kidney; a separate classification is published for nephroblastomas.

HISTOPATHOLOGIC GRADE (G)

GX Grade cannot be assessed
G1 Well differentiated
G2 Moderately differentiated
G3-4 Poorly differentiated of undifferentiated

BIBLIOGRAPHY

1. Angervall L, Carlstrom E, Wahlquist L, et al: Effects of clinical and morphological variables on spread of renal carcinoma in an operative series. Scand J Urol Nephrol 3:134–140, 1969
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10. Weyman PJ, McClennan BL, Stanley RJ, et al: Comparison of computer tomography and angiography in the evaluation of renal carcinoma. Radiology 137:417–424, 1980

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age ____ Sex ____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

DEFINITIONS

Primary Tumor (T)

Clin	Path		
[ ]	[ ]	TX	Primary tumor cannot be assessed
[ ]	[ ]	T0	No evidence of primary tumor
[ ]	[ ]	T1	Tumor 2.5 cm or less in greatest dimension limited to the kidney
[ ]	[ ]	T2	Tumor more than 2.5 cm in greatest dimension limited to the kidney
[ ]	[ ]	T3	Tumor extends into major veins or invades adrenal gland or perinephric tissues but not beyond Gerota's fascia
[ ]	[ ]	T3a	Tumor invades adrenal gland or perinephric tissues but not beyond Gerota's fascia
[ ]	[ ]	T3b	Tumor grossly extends into renal vein(s) or vena cava below diaphragm
[ ]	[ ]	T3c	Tumor grossly extends into vena cava above diaphragm
[ ]	[ ]	T4	Tumor invades beyond Gerota's fascia

Lymph Node (N)

Clin	Path		
[ ]	[ ]	NX	Regional lymph nodes cannot be assessed
[ ]	[ ]	N0	No regional lymph node metastasis
[ ]	[ ]	N1	Metastasis in a single lymph node, 2 cm or less in greatest dimension
[ ]	[ ]	N2	Metastasis in a single lymph node, more than 2 cm but not more than 5 cm in greatest dimension, or multiple lymph nodes, none more than 5 cm in greatest dimension
[ ]	[ ]	N3	Metastasis in a lymph node more than 5 cm in greatest dimension

Distant Metastasis (M)

Clin	Path		
[ ]	[ ]	MX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0	No distant metastasis
[ ]	[ ]	M1	Distant metastasis

Stage Grouping

Clin	Path				
[ ]	[ ]	I	T1	N0	M0
[ ]	[ ]	II	T2	N0	M0
[ ]	[ ]	III	T1	N1	M0
			T2	N1	M0
			T3a	N0	M0
			T3a	N1	M0
			T3b	N0	M0
			T3b	N1	M0
			T3c	N0	M0
			T3c	N1	M0
[ ]	[ ]	IV	T4	Any N	M0
			Any T	N2	M0
			Any T	N3	M0
			Any T	Any N	M1

Staged by _____ M.D.
_____ Registrar
Date _____

(continued on next page)

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated
[ ] G3-4 Poorly differentiated or undifferentiated

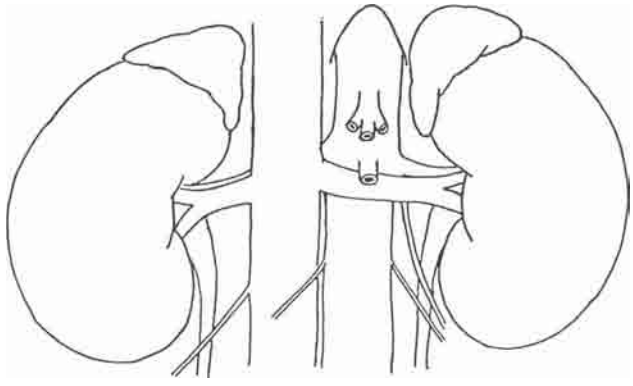
Histopathologic Type

The histopathologic types are:

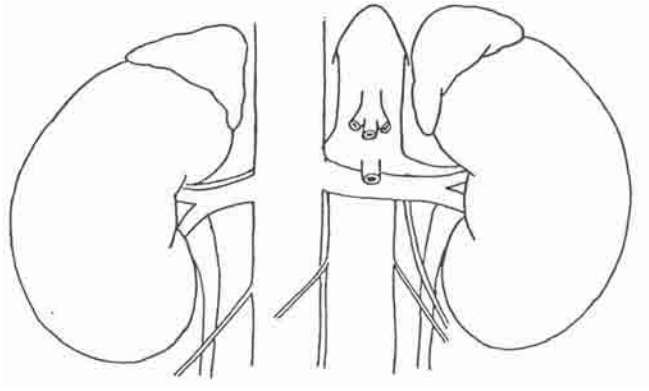
Renal cell carcinoma
Adenocarcinoma
Renal papillary adenocarcinoma
Tubular carcinoma
Granular cell carcinoma
Clear cell carcinoma—Hypernephroma

The predominant cancer is adenocarcinoma; subtypes are clear-cell and granular-cell carcinoma. A grading system is recommended when feasible. The staging system does not apply to sarcomas of the kidney. A separate classification is published for nephroblastomas.

Illustrations



This drawing is to be used with the checklist. Sketch in the urographic, angiographic, ultrasound, or CT extent of tumor.



This drawing is to be used with the checklist and the upper drawing. Sketch in the pathologic extent of tumor.

36

Renal Pelvis and Ureter

C65.9 Renal pelvis
C66.9 Ureter

Tumors of the renal pelvis and ureter are not common. Tumors of the renal pelvis comprise only 5% to 10% of all renal cancers. Most cases are found in adults. Commonly, malignant tumors in the renal pelvis or ureter are multiple and associated with cancers located in other parts of the urinary tract. For instance, carcinomas of the ureter are often associated with tumors in the urinary bladder. Most tumors are transitional cell carcinomas, although other types can occur. Tumors in the renal pelvis may be associated with calculi. Staging depends on the extent of disease. T3 differs between the renal pelvis and the ureter, but all other definitions are the same.

ANATOMY

Primary Site. The renal pelvis and ureter form a single unit that cephalad is continuous with the collecting ducts of the renal pyrimides and comprises the minor and major calyces, which are continuous with the renal pelvis. The ureteropelvic junction is variable in position and location, but serves as a "landmark" that separates the renal pelvis and the ureter, which continues caudad and traverses the wall of the urinary bladder as the intramural ureter opening on the trigone of the bladder at the ureteral orifice. The renal pelvis and ureter are composed of the following layers: epithelium, subepithelial connective tissue, and muscularis, which is continuous with a connective tissue adventitial layer. It is in this outer layer that the major blood supply and lymphatics are found.

The intrarenal portion of the renal pelvis is surrounded by renal parenchyma; the extrarenal pelvis, by perihilar fat. The ureter courses through the retroperitoneum adjacent to the parietal peritoneum and rests on the retroperitoneal musculature above the pelvic vessels. As it crosses the vessels and enters the deep pelvis, the ureter is surrounded by pelvic fat until it traverses the bladder wall.

Regional Lymph Nodes. The regional lymph nodes include:

Renal pelvis:
- Renal hilar
- Paracaval
- Aortic
- Retroperitoneal, NOS

Ureter:
- Renal hilar
- Iliac (common, internal hypogastric, external)
- Paracaval
- Periureteral
- Pelvic, NOS

The significance of regional lymph node metastasis in staging renal cancer lies in the number and size and not in whether metastasis is unilateral or contralateral.

Metastatic Sites. Distant spread to lung, bone, and liver is most common.

RULES FOR CLASSIFICATION

Clinical Staging. Primary tumor assessment includes radiographic imaging, endoscopic evaluation, and ureteroscopy when applicable. Material in cytoscopic study should be obtained. The possible concurrent presence of bladder tumors is not a prognostic factor in this staging system. These tumors should be staged separately. Evaluation of distant metastatic sites includes radiographic, radioisotopic, and appropriate blood studies.

Pathologic Staging. Histologic confirmation of extent of disease is required. Resection of primary tumor, kidney, ureter, appropriate regional lymph nodes, and bladder cuff is usually required. Special circumstances may limit the magnitude of resection, but, at a minimum, the tumor with appropriate margins and regional lymph

nodes must be available for pathologic evaluation. Laterality does not affect N classification.

DEFINITION OF TNM

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
Ta Papillary noninvasive carcinoma
Tis Carcinoma *in situ*
T1 Tumor invades subepithelial connective tissue
T2 Tumor invades the muscularis
T3 (For renal pelvis only) Tumor invades beyond the muscularis into peripelvic fat or the renal parenchyma
T3 (For ureter only) Tumor invades beyond the muscularis into periureteric fat
T4 Tumor invades adjacent organs, or through the kidney into the parinephric fat.

Regional Lymph Nodes (N)*

NX Regional lymph nodes cannot be assessed
N0 No regional lymph node metastasis
N1 Metastasis in a single lymph node, 2 cm or less in greatest dimension
N2 Metastasis in a single lymph node, more than 2 cm but not more than 5 cm in greatest dimension; or multiple lymph nodes, none more than 5 cm in greatest dimension
N3 Metastasis in a lymph node more than 5 cm in greatest dimension

* *Note:* Laterality does not affect N classification.

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

STAGE GROUPING

Stage 0a	Ta	N0	M0
Stage 0is	Tis	N0	M0
Stage I	T1	N0	M0
Stage II	T2	N0	M0
Stage III	T3	N0	M0
Stage IV	T4	N0	M0
	Any T	N1	M0
	Any T	N2	M0
	Any T	N3	M0
	Any T	Any N	M1

HISTOPATHOLOGIC TYPE

The histologic types are:

Transitional cell carcinoma
Papillary carcinoma
Squamous cell carcinoma
Epidermoid carcinoma
Adenocarcinoma
Urothelial carcinoma

HISTOPATHOLOGIC GRADE

GX Grade cannot be assessed
G1 Well differentiated
G2 Moderately differentiated
G3-4 Poorly differentiated or undifferentiated

BIBLIOGRAPHY

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Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	DEFINITIONS
		Primary Tumor (T)
[ ]	[ ]	TX Primary tumor cannot be assessed
[ ]	[ ]	T0 No evidence of primary tumor
[ ]	[ ]	Ta Papillary noninvasive carcinoma
[ ]	[ ]	Tis Carcinoma *in situ*
[ ]	[ ]	T1 Tumor invades subepithelial connective tissue
[ ]	[ ]	T2 Tumor invades muscularis ~~(For ureter only)~~ a
[ ]	[ ]	T3 Tumor invades beyond muscularis into peripelvic fat or renal parenchyma ~~(For ureter only)~~ (For renal pelvis only) b
[ ]	[ ]	T3 Tumor invades beyond muscularis into peripelvic fat (For ureter only) c
[ ]	[ ]	T4 Tumor invades adjacent organs or through the kidney into parinephric fat
		Lymph Node (N)
[ ]	[ ]	NX Regional lymph nodes cannot be assessed
[ ]	[ ]	N0 No regional lymph node metastasis
[ ]	[ ]	N1 Metastasis in a single lymph node, 2 cm or less in greatest dimension
[ ]	[ ]	N2 Metastasis in a single lymph node, more than 2 cm but not more than 5 cm in greatest dimension, or multiple lymph nodes, none more than 5 cm in greatest dimension
[ ]	[ ]	N3 Metastasis in a lymph node more than 5 cm in greatest dimension
		Distant Metastasis (M)
[ ]	[ ]	MX Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0 No distant metastasis
[ ]	[ ]	M1 Distant metastasis

Clin	Path	Stage Grouping			
[ ]	[ ]	0a	Ta	N0	M0
[ ]	[ ]	0is	Tis	N0	M0
[ ]	[ ]	I	T1	N0	M0
[ ]	[ ]	II	T2	N0	M0
[ ]	[ ]	III	T3	N0	M0
[ ]	[ ]	IV	T4	N0	M0
			Any T	N1	M0
			Any T	N2	M0
			Any T	N3	M0
			Any T	Any N	M1

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated
[ ] G3-4 Poorly differentiated or undifferentiated

Histopathologic Type

The histopathologic types are:

Transitional cell carcinoma
Papillary carcinoma
Squamous cell carcinoma
Epidermoid carcinoma
Adenocarcinoma
Urothelial carcinoma

Staged by _____ M.D.
_____ Registrar
Date _____

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Urethra

C68.0 Urethra
C68.1 Paraurethral gland
C68.8 Overlapping lesion
C68.9 Urinary system, NOS

In both sexes, cancers of the urethra are exceedingly rare. Most carcinomas of the female urethra occur at the junction of the transitional and stratified squamous epithelium at the meatus. In males, the cancer may be associated with a venereal disease, such as gonorrhea; the most common location is the bulbomembranous portion. In females and males, most tumors are squamous cell carcinomas. In males, transitional cell carcinomas are found in the prostatic portion. Staging depends on the depth of penetration and local extension.

ANATOMY

Primary Site. In the *male*, the urethra is divided into anterior, penile (pendulous), and posterior (bulbomembranous and prostate). The urethra consists of mucosa, submucosal stroma, and the surrounding corpus spongiosum. Histologically, the meatal and parameatal urethra are lined with squamous epithelium; the penile and bulbomembranous urethra, with pseudostratified or stratified columnar epithelium; and the prostatic urethra, with transitional cell epithelium. The corpora cavernosum is contiguous to the bulbous and penile urethra.

The *female* urethra is divided into proximal and distal sections. The epithelium is supported on subepithelial connective tissue. The periurethral glands of Skene are concentrated near the meatus but extend along the entire urethra. The urethra is surrounded by a longitudinal layer of smooth muscle continuous with the bladder. The distal third of the urethra is contiguous to the vaginal wall. The distal two-thirds of the urethra is lined with squamous epithelium; the proximal one third, with transitional epithelium. The periurethral glands are lined with pseudostratified and stratified columnar epithelium.

Regional Lymph Nodes. The regional lymph nodes include:

Iliac (common, internal [hypogastric] obturator, external)
Inguinal (superficial or deep)
Presacral
Sacral, NOS
Pelvic, NOS

The significance of regional lymph node metastasis in staging urethral cancer lies in the number and size and not in whether metastasis is unilateral or bilateral.

Metastatic Sites. Distant spread to lung, liver, and bone is most common.

RULES FOR CLASSIFICATION

Clinical Staging. Radiographic imaging, cystourethroscopy, palpation, and biopsy or cytology of the tumor prior to definitive treatment are desirable. The site of origin should be confirmed to exclude metastatic disease.

Pathologic Staging. Histologic examination and confirmation of extent and location of disease are required. The extent of resection, including removal of regional lymph nodes, will depend on tumor location, depth of penetration, and sex of patient. Laterality does not affect the N classification.

DEFINITION OF TNM

Primary Tumor (T) (male and female)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
Ta Noninvasive papillary, polypoid, or verrucous carcinoma

Tis Carcinoma *in situ*
T1 Tumor invades subepithelial connective tissue
T2 Tumor invades the corpus spongiosum or the prostate, or the periurethral muscle
T3 Tumor invades the corpus cavernosum or beyond the prostatic capsule, or the anterior vagina or bladder neck
T4 Tumor invades other adjacent organs

Regional Lymph Nodes (N)

NX Regional lymph nodes cannot be assessed
N0 No regional lymph node metastasis
N1 Metastasis in a single lymph node, 2 cm or less in greatest dimension
N2 Metastasis in a single lymph node, more than 2 cm but not more than 5 cm in greatest dimension; or multiple lymph nodes, none more than 5 cm in greatest dimension
N3 Metastasis in a lymph node more than 5 cm in greatest dimension

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

STAGE GROUPING

Stage 0a	Ta	N0	M0
Stage 0is	Tis	N0	M0
Stage I	T1	N0	M0
Stage II	T2	N0	M0
Stage III	T1	N1	M0
	T2	N1	M0
	T3	N0	M0
	T3	N1	M0
Stage IV	T4	N0	M0
	T4	N1	M0
	Any T	N2	M0
	Any T	N3	M0
	Any T	Any N	M1

HISTOPATHOLOGIC TYPE

Cell types can be divided into transitional, squamous, and glandular.

HISTOPATHOLOGIC GRADE (G)

GX Grade cannot be assessed
G1 Well differentiated
G2 Moderately differentiated
G3-4 Poorly differentiated or undifferentiated

BIBLIOGRAPHY

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3. Vernon HK, Wilkins RD: Primary carcinoma of the male urethra. Br J Urol 21:232–235, 1950

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

DEFINITIONS

Primary Tumor (T)

Clin	Path		
[ ]	[ ]	TX	Primary tumor cannot be assessed
[ ]	[ ]	T0	No evidence of primary tumor
[ ]	[ ]	Ta	Noninvasive papillary, polypoid, or verrucous carcinoma
[ ]	[ ]	Tis	Carcinoma *in situ*
[ ]	[ ]	T1	Tumor invades subepithelial connective tissue
[ ]	[ ]	T2	Tumor invades corpus spongiosum or prostate or periurethral muscle
[ ]	[ ]	T3	Tumor invades corpus cavernosum or beyond prostatic capsule or the anterior vagina or bladder neck
[ ]	[ ]	T4	Tumor invades other adjacent organs

Lymph Node (N)

[ ]	[ ]	NX	Regional lymph nodes cannot be assessed
[ ]	[ ]	N0	No regional lymph node metastasis
[ ]	[ ]	N1	Metastasis in a single lymph node, 2 cm or less in greatest dimension
[ ]	[ ]	N2	Metastasis in a single lymph node, more than 2 cm but not more than 5 cm in greatest dimension, or multiple lymph nodes, none more than 5 cm in greatest dimension
[ ]	[ ]	N3	Metastasis in a lymph node more than 5 cm in greatest dimension

Distant Metastasis (M)

[ ]	[ ]	MX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0	No distant metastasis
[ ]	[ ]	M1	Distant metastasis

Stage Grouping

Clin	Path				
[ ]	[ ]	0a	Ta	N0	M0
[ ]	[ ]	0is	Tis	N0	M0
[ ]	[ ]	I	T1	N0	M0
[ ]	[ ]	II	T2	N0	M0
[ ]	[ ]	III	T1	N1	M0
			T2	N1	M0
			T3	N0	M0
			T3	N1	M0
[ ]	[ ]	IV	T4	N0	M0
			T4	N1	M0
			Any T	N2	M0
			Any T	N3	M0
			Any T	Any N	M1

Histopathologic Type

Cell types can be divided into transitional, squamous, and glandular.

Histopathologic Grade (G)

[ ]	GX	Grade cannot be assessed
[ ]	G1	Well differentiated
[ ]	G2	Moderately differentiated
[ ]	G3-4	Poorly differentiated or undifferentiated

Staged by _____ M.D.
_____ Registrar
Date _____

OPHTHALMIC CANCERS

The orbit and its contents—primarily the eye—contain many types of tissues. Consequently, a wide variety of malignant tumors occur in this anatomic area. This section includes recommendations for staging these cancers based on data available in the literature and on knowledge of the experts serving on the American Joint Committee on Cancer's Task Force for Staging of Cancer of the Eye.

The following sites are included:

Eyelid
Conjunctiva
Uvea
Retina
Orbit
Lacrimal gland

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Carcinoma of the Eyelid

C44.1 Eyelid

ANATOMY

Primary Site. The eyelid is covered externally by epidermis and internally by conjunctiva, which becomes continuous with the conjunctiva covering the eyeball. Basal cell carcinomas and squamous cell carcinomas arise from the epidermal surface. Sebaceous cell carcinomas arise from the meibomian glands in the tarsus, the glands of Zeis at the lid margin, and the sebaceous glands of the caruncle. Other adnexal carcinomas arise from the sweat glands of Moll and the hair follicles.

Regional Lymph Nodes. The eyelids are supplied with lymphatics that drain into the preauricular, infra-auricular, facial, submandibular, and cervical lymph nodes.

Metastatic Sites. Tumors of the eyelids not only metastasize to distant sites by way of the regional lymphatics and bloodstream but also spread directly into the orbit, including the lacrimal gland, and into the eyeball.

RULES FOR CLASSIFICATION

The classification applies only to carcinoma. There should be histologic verification of the cancer. This verification permits a division of cases by histologic type (i.e., basal cell, squamous cell, and sebaceous carcinoma). Any unconfirmed case must be reported separately.

Clinical Staging. The assessment of the cancer is based on inspection, slit-lamp examination, palpation of the regional lymph nodes, and, when indicated, radiologic (including computed tomography) and ultrasonographic examination of the orbit, paranasal sinuses, and chest.

Pathologic Staging. Complete resection of the primary site is indicated. Histologic study of the margins and the deep aspect of resected tissues is necessary. Resection or needle biopsy of enlarged regional lymph nodes or orbital masses is desirable.

DEFINITION OF TNM

The following definitions apply to both clinical and pathologic staging.

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor

Tis	Carcinoma *in situ*
T1	Tumor of any size, not invading the tarsal plate or, at the eyelid margin, 5 mm or less in greatest dimension
T2	Tumor invades tarsal plate or, at the eyelid margin, more than 5 mm but not more than 10 mm in greatest dimension
T3	Tumor involves full eyelid thickness or, at the eyelid margin, more than 10 mm in greatest dimension
T4	Tumor invades adjacent structures

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

MX	Presence of distant metastasis cannot be assessed
M0	No distant metastasis
M1	Distant metastasis

STAGE GROUPING

No stage grouping is presently recommended.

HISTOPATHOLOGIC TYPE

Basal cell carcinoma
Squamous cell carcinoma
Sebaceous cell carcinoma

HISTOPATHOLOGIC GRADE (G)

GX	Grade cannot be assessed
G1	Well differentiated
G2	Moderately differentiated
G3	Poorly differentiated
G4	Undifferentiated

BIBLIOGRAPHY

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Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	DEFINITIONS	
		Primary Tumor (T)	
[ ]	[ ]	TX	Primary tumor cannot be assessed
[ ]	[ ]	T0	No evidence of primary tumor
[ ]	[ ]	Tis	Carcinoma *in situ*
[ ]	[ ]	T1	**Tumor of any size**, not invading the tarsal plate or, at the eyelid margin, 5 mm or less in greatest dimension
[ ]	[ ]	T2	Tumor invades tarsal plate or, at the eyelid margin, more than 5 mm but not more than 10 mm in greatest dimension
[ ]	[ ]	T3	Tumor involves full eyelid thickness or, at the eyelid margin, more than 10 mm in greatest dimension
[ ]	[ ]	T4	Tumor invades adjacent structures
		Lymph Node (N)	
[ ]	[ ]	NX	Regional lymph nodes cannot be assessed
[ ]	[ ]	N0	No regional lymph node metastasis
[ ]	[ ]	N1	Regional lymph node metastasis
		Distant Metastasis (M)	
[ ]	[ ]	MX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0	No distant metastasis
[ ]	[ ]	M1	Distant metastasis

Stage Grouping

No stage grouping is presently recommended.

Staged by _____ M.D.
_____ Registrar
Date _____

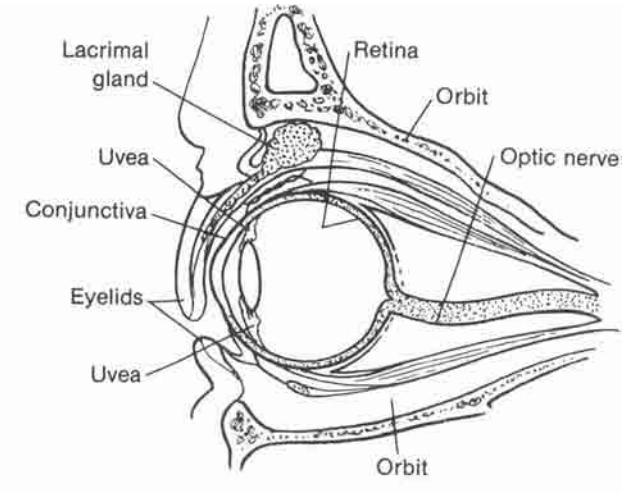
Histopathologic Type

Basal cell carcinoma
Squamous cell carcinoma
Sebaceous carcinoma

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated
[ ] G3 Poorly differentiated
[ ] G4 Undifferentiated

Illustration



Indicate on diagram and describe exact location and characteristics of tumor.

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Malignant Melanoma of the Eyelid

C44.1 Eyelid

This chapter has been adapted from the discussion for melanoma of the skin, because that discussion is considered applicable to melanoma of the skin of the eyelid.

No cT categories are presently recommended.

The pT categories correspond to those in the third edition of the AJCC manual and are based on Clark's "levels" and Breslow's "thickness of invasion." Thickness of invasion into the skin is recorded as an actual measurement by an ocular micrometer.

Maximal thickness of the tumor is measured with an ocular micrometer at a right angle to the adjacent normal skin. The upper reference point is the top of the granular cell layer of the epidermis of the overlying skin, or the base of the lesion if the tumor is ulcerated. The lower reference point is usually the deepest point of invasion. It may be the invading edge of a single tumor mass or an isolated cell or group of cells deep to the main mass.

The N and M categories correspond to those of melanoma of the skin.

ANATOMY

Primary Site. The eyelid is covered externally by epidermis and internally by conjunctiva, which becomes continuous with the conjunctiva covering the eyeball.

Regional Lymph Nodes. The eyelids are supplied with lymphatics that drain into the preauricular, infra-auricular, facial, submandibular, and cervical lymph nodes.

Metastatic Sites. Tumors of the eyelid not only metastasize to distant sites by way of the regional lymphatics and bloodstream but also spread directly into the orbit, including the lacrimal gland, and into the eyeball.

RULES FOR CLASSIFICATION

Clinical Staging. Assessment of the cancer is based on inspection, slit-lamp examination, palpation of the regional lymph nodes, and, when indicated, radiologic (including computed tomography) and ultrasonographic examination of the orbit, paranasal sinuses, and chest.

Pathologic Staging. Complete resection of the primary site is indicated. Histologic study of the margins and the deep aspect of resected tissues is necessary. Resection or needle biopsy of enlarged regional lymph nodes or orbital masses is desirable.

DEFINITION OF TNM

Clinical Classification (cTNM)

Primary Tumor (T)

No classification is recommended at present.

Regional Lymph Nodes (N)

- NX Regional lymph nodes cannot be assessed
- N0 No regional lymph node metastasis
- N1 Metastasis 3 cm or less in greatest dimension in any regional lymph node(s)
- N2 Metastasis more than 3 cm in greatest dimension in any regional lymph node(s) and/or in-transit metastasis
  - N2a Metastasis more than 3 cm in greatest dimension in any regional node(s)
  - N2b In-transit metastasis
  - N2c Both (N2a and N2b)

Distant Metastasis (M)

- MX Presence of distant metastasis cannot be assessed
- M0 No distant metastasis
- M1 Distant metastasis

DEFINITION OF TNM

Pathologic Classification (pTNM)

Primary Tumor (pT)

pTX Primary tumor cannot be assessed
pT0 No evidence of primary tumor
pTis Melanoma *in situ* (atypical melanocytic hyperplasia, severe melanocytic dysplasia), not an invasive malignant lesion (Clark's Level I)
pT1 Tumor 0.75 mm or less in thickness and invades the papillary dermis (Clark's Level II)
pT2 Tumor more than 0.75 mm but not more than 1.5 mm in thickness and/or invades to the papillary-reticular dermal interface (Clark's Level III)
pT3 Tumor more than 1.5 mm but not more than 4 mm in thickness and/or invades the reticular dermis (Clark's Level IV)
- pT3a Tumor more than 1.5 mm but not more than 3 mm in thickness
- pT3b Tumor more than 3 mm but not more than 4 mm in thickness

pT4 Tumor more than 4 mm in thickness and/or invades the subcutaneous tissue and/or satellite(s) within 2 cm of the primary tumor (Clark's Level V)
- pT4a Tumor more than 4 mm in thickness and/or invades the subcutaneous tissue
- pT4b Satellite(s) within 2 cm of the primary tumor

Regional Lymph Nodes (pN)

pNX Regional lymph nodes cannot be assessed
pN0 No regional lymph node metastasis
pN1 Metastasis 3 cm or less in greatest dimension in any regional lymph node(s)
pN2 Metastasis more than 3 cm in greatest dimension in any regional lymph node(s) and/or in-transit metastasis
- pN2a Metastasis more than 3 cm in greatest dimension
- pN2b In-transit metastasis
- pN2c Both (pN2a and pN2b)

Distant Metastasis (pM)

pMX Presence of distant metastasis cannot be assessed
pM0 No distant metastasis
pM1 Distant metastasis
- pM1a Metastasis in skin or subcutaneous tissue or lymph node(s) beyond the regional lymph nodes
- pM1b Visceral metastasis

Note: In-transit metastasis involves skin or subcutaneous tissue more than 2 cm from the primary tumor not beyond the regional lymph nodes.

STAGE GROUPING

Stage I	pT1	N0	M0
	pT2	N0	M0
Stage II	pT3	N0	M0
Stage III	pT4	N0	M0
	Any pT	N1	M0
	Any pT	N2	M0
Stage IV	Any pT	Any N	M1

HISTOPATHOLOGIC TYPE

This classification is only for melanoma of the eyelid.

BIBLIOGRAPHY

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Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path		
		DEFINITIONS	
		Primary Tumor (T)	
		No classification is recommended at present.	
		Lymph Node (N)	
[ ]	[ ]	NX	Regional lymph nodes cannot be assessed
[ ]	[ ]	N0	No regional lymph node metastasis
[ ]	[ ]	N1	Metastasis 3 cm or less in greatest dimension in any regional lymph node(s)
[ ]	[ ]	N2	Metastasis more than 3 cm in greatest dimension in any regional lymph node(s) and/or in-transit metastasis
[ ]	[ ]	N2a	Metastasis more than 3 cm in greatest dimension in any regional node(s)
[ ]	[ ]	N2b	In-transit metastasis
[ ]	[ ]	N2c	Both
		Primary Tumor (pT)	
[ ]	[ ]	pTX	Primary tumor cannot be assessed
[ ]	[ ]	pT0	No evidence of primary tumor
[ ]	[ ]	pTis	Melanoma *in situ* (atypical melanocytic hyperplasia, severe melanocytic dysplasia), not an invasive malignant lesion (Clark's Level I)
[ ]	[ ]	pT1	Tumor 0.75 mm or less in thickness and invades the papillary dermis (Clark's Level II)
[ ]	[ ]	pT2	Tumor more than 0.75 mm but not more than 1.5 mm in thickness and/or invades to the papillary-reticular dermal interface (Clark's Level III)
[ ]	[ ]	pT3	Tumor more than 1.5 mm but not more than 4 mm in thickness and/or invades the reticular dermis (Clark's Level IV)
[ ]	[ ]	pT3a	Tumor more than 1.5 mm but not more than 3 mm in thickness
[ ]	[ ]	pT3b	Tumor more than 3 mm but not more than 4 mm in thickness
[ ]	[ ]	pT4	Tumor more than 4 mm in thickness and/or invades the subcutaneous tissue and/or satellite(s) within 2 cm of the primary tumor (Clark's Level V)
[ ]	[ ]	pT4a	Tumor more than 4 mm in thickness and/or invades the subcutaneous tissue
[ ]	[ ]	pT4b	Satellite(s) within 2 cm of the primary tumor
		Lymph Node (pN)	
[ ]	[ ]	pNX	Regional lymph nodes cannot be assessed
[ ]	[ ]	pN0	No regional lymph node metastasis
[ ]	[ ]	pN1	Metastasis 3 cm or less in greatest dimension in any regional lymph node(s)
[ ]	[ ]	pN2	Metastasis more than 3 cm in greatest dimension in any regional lymph node(s) and/or in-transit metastasis
[ ]	[ ]	pN2a	Metastasis more than 3 cm in greatest dimension
[ ]	[ ]	pN2b	In-transit metastasis
[ ]	[ ]	pN2c	Both (pN2a and pN2b)
		Distant Metastasis (pM)	
[ ]	[ ]	pMX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	pM0	No distant metastasis
[ ]	[ ]	pM1	Distant metastasis
[ ]	[ ]	pM1a	Metastasis in skin or subcutaneous tissue or lymph node(s) beyond the regional lymph nodes
[ ]	[ ]	pM1b	Visceral metastasis

(continued on next page)

Clin	Path
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]

Stage Grouping

I	pT1	N0	M0
	pT2	N0	M0
II	pT3	N0	M0
III	pT4	N0	M0
	Any pT	N1	M0
IV	Any pT	Any N	M1

Staged by _____ M.D.

_____ Registrar

Date _____

Histopathologic Type

This classification is only for melanoma of the eyelid.

Note: In-transit metastasis involves skin or subcutaneous tissue more than 2 cm from the primary tumor not beyond the regional lymph nodes.

40

Carcinoma of the Conjunctiva

C69.0 Conjunctiva

ANATOMY

Primary Site. The conjunctiva consists of stratified epithelium that contains mucus-secreting goblet cells; these cells are most numerous in the fornices. Palpebral conjunctiva lines the eyelid; bulbar conjunctiva covers the eyeball. Conjunctival epithelium merges with that of the cornea at the limbus. It is at this site, particularly at the temporal limbus, that carcinoma is most likely to arise. Conjunctival intraepithelial neoplasia (CIN) embraces all forms of intraepithelial dysplasia, including *in situ* carcinoma. Mucinous adenocarcinoma is a rare form of adenocarcinoma of the conjunctival goblet cells.

Regional Lymph Nodes. The regional lymph nodes are:

Preauricular (parotid)
Submandibular
Cervical

Metastatic Sites. Tumors of the conjunctiva, in addition to spread by way of regional lymphatics, may also involve the eyelid proper, the orbit, lacrimal glands, and the brain.

RULES FOR CLASSIFICATION

Clinical Staging. Assessment of the cancer is based on inspection, slit-lamp examination, palpation of the regional lymph nodes, and, when indicated, radiologic examination (including computed tomography) and ultrasonographic examination of the orbit, paranasal sinuses, and chest.

Pathologic Staging. Complete resection of the primary site is indicated. Extensive local involvement of orbital spread requires exenteration. Histologic study of the margins of the deep aspect of resected tissues is necessary.

DEFINITION OF TNM

These definitions apply to both clinical and pathologic staging.

Primary Tumor (T)

TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
Tis	Carcinoma *in situ*
T1	Tumor 5 mm or less in greatest dimension
T2	Tumor more than 5 mm in greatest dimension, without invasion of adjacent structures
T3	Tumor invades adjacent structures, excluding the orbit
T4	Tumor invades the orbit

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

MX	Presence of distant metastasis cannot be assessed
M0	No distant metastasis
M1	Distant metastasis

STAGE GROUPING

No stage grouping is presently recommended.

HISTOPATHOLOGIC TYPE

This classification applies only to carcinoma of the conjunctiva.

HISTOPATHOLOGIC GRADE (G)

GX	Grade cannot be assessed
G1	Well differentiated
G2	Moderately differentiated
G3	Poorly differentiated
G4	Undifferentiated

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age ____ Sex ____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

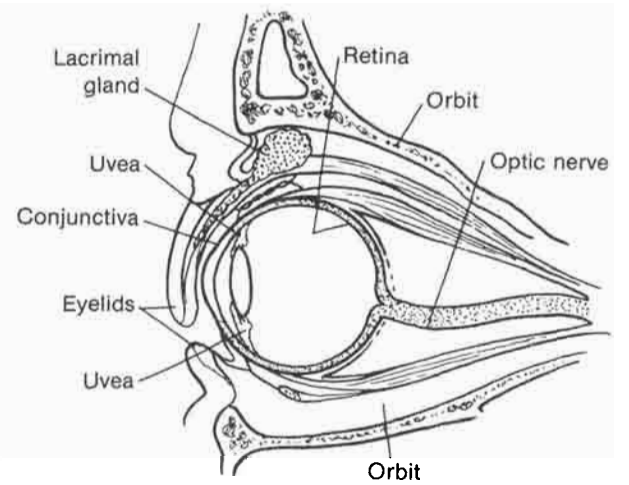
Clin	Path	DEFINITIONS	
		Primary Tumor (T)	
[ ]	[ ]	TX	Primary tumor cannot be assessed
[ ]	[ ]	T0	No evidence of primary tumor
[ ]	[ ]	Tis	Tumor *in situ*
[ ]	[ ]	T1	**Tumor 5 mm** or less in greatest dimension
[ ]	[ ]	T2	Tumor more than 5 mm in greatest dimension, without invasion of adjacent structures
[ ]	[ ]	T3	Tumor invades adjacent structures, excluding the orbit
[ ]	[ ]	T4	Tumor invades the orbit
		Lymph Node (N)	
[ ]	[ ]	NX	Regional lymph nodes cannot be assessed
[ ]	[ ]	N0	No regional lymph node metastasis
[ ]	[ ]	N1	Regional lymph node metastasis
		Distant Metastasis (M)	
[ ]	[ ]	MX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0	No distant metastasis
[ ]	[ ]	M1	Distant metastasis

Stage Grouping

No stage grouping is presently recommended.

Staged by _____ M.D.
_____ Registrar
Date _____

Illustration



Indicate on diagram and describe exact location and characteristics of tumor.

Histopathologic Type

This classification applies only to carcinoma of conjunctiva.

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated
[ ] G3 Poorly differentiated
[ ] G4 Undifferentiated

41

Malignant Melanoma of the Conjunctiva

C69.0 Conjunctiva

ANATOMY

Primary Site. In addition to mucus-secreting goblet cells within the stratified epithelium, melanocytic cells exist in the basal layer. These are of neuroectodermal origin, and melanocytic tumors may arise from these cells. Melanomas may arise from junctional and compound nevi, from primary acquired melanosis, or de novo. Tumors must be distinguished from nontumorous pigmentation.

Regional Lymph Nodes. The regional lymph nodes are:

Parotid
Preauricular
Submandibular
Cervical

Metastatic Sites. In addition to spread by lymphatics and the bloodstream, direct extension to the eyeball and orbit occurs.

RULES FOR CLASSIFICATION

The classification applies only to melanoma. There should be histologic verification of the melanocytic lesion.

Clinical Staging. The assessment of the cancer is based on inspection, slit-lamp examination, palpation of the regional lymph nodes, and, when indicated, radiologic (including computed tomography) and ultrasonographic examination of the orbit, paranasal sinuses, and chest.

Pathologic Staging. Complete resection of the primary site is indicated. Histologic study of the margins and the deep aspect of resected tissues is necessary. Resection or needle biopsy of enlarged regional lymph nodes or orbital masses is desirable.

DEFINITION OF TNM

Clinical Classification

Primary Tumor (T)

TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
T1	Tumor(s) of the bulbar conjunctiva occupying one quadrant or less
T2	Tumor(s) of the bulbar conjunctiva occupying more than one quadrant
T3	Tumor(s) of the conjunctival fornix and/or palpebral conjunctiva and/or caruncle
T4	Tumor invades the eyelid, cornea, and/or orbit

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

MX	Presence of distant metastasis cannot be assessed
M0	No distant metastasis
M1	Distant metastasis

Pathologic Classification (pTNM)

Primary Tumor (pT)

pTX	Primary tumor cannot be assessed
pT0	No evidence of primary tumor
pT1	Tumor(s) of the bulbar conjunctiva occupying one quadrant or less and 2 mm or less in thickness

pT2 Tumor(s) of the bulbar conjunctiva occupying more than one quadrant and 2 mm or less in thickness
pT3 Tumor(s) of the conjunctival fornix and/or palpebral conjunctiva and/or caruncle or tumor(s) of the bulbar conjunctiva, more than 2 mm in thickness
pT4 Tumor invades the eyelid, cornea, and/or orbit

Regional Lymph Nodes (pN)

pNX Regional lymph nodes cannot be assessed
pN0 No regional lymph node metastasis
pN1 Regional lymph node metastasis

Distant Metastasis (pM)

pMX Presence of distant metastasis cannot be assessed
pM0 No distant metastasis
pM1 Distant metastasis

STAGE GROUPING

No stage grouping is presently recommended.

HISTOPATHOLOGIC TYPE

This categorization applies only to melanoma of the conjunctiva.

HISTOPATHOLOGIC GRADE (G)

Histopathologic grade represents the origin of the primary tumor.

GX Origin cannot be assessed
G0 Primary acquired melanosis
G1 Malignant melanoma arises from a nevus
G2 Malignant melanoma arises from primary acquired melanosis
G3 Malignant melanoma arises de novo

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3. Zimmerman LE: The histogenesis of conjunctival melanomas: The first Algernon B. Reese lecture. In Jakobiec FA (Ed.), Ocular and Adnexal Tumors. Birmingham, Aesculapius, 1978

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path		
		DEFINITIONS	
		Primary Tumor (T)	
[ ]	[ ]	TX	Primary tumor cannot be assessed
[ ]	[ ]	T0	No evidence of primary tumor
[ ]	[ ]	T1	Tumor(s) of bulbar conjunctiva occupying one quadrant or less
[ ]	[ ]	T2	Tumor(s) of bulbar conjunctiva occupying more than one quadrant
[ ]	[ ]	T3	Tumor(s) of conjunctival fornix and/or palpebral conjunctiva and/or caruncle
[ ]	[ ]	T4	Tumor invades eyelid, cornea, and/or orbit
		Lymph Node (N)	
[ ]	[ ]	NX	Regional lymph nodes cannot be assessed
[ ]	[ ]	N0	No regional lymph node metastasis
[ ]	[ ]	N1	Regional lymph node metastasis
		Distant Metastasis (M)	
[ ]	[ ]	MX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0	No distant metastasis
[ ]	[ ]	M1	Distant metastasis
		Primary Tumor (pT)	
[ ]	[ ]	pTX	Primary tumor cannot be assessed
[ ]	[ ]	pT0	No evidence of primary tumor
[ ]	[ ]	pT1	Tumor(s) of bulbar conjunctiva occupying one quadrant or less and 2 mm or less in thickness
[ ]	[ ]	pT2	Tumor(s) of bulbar conjunctiva occupying more than one quadrant and 2 mm or less in thickness
[ ]	[ ]	pT3	Tumor(s) of conjunctival fornix and/or palpebral conjunctiva and/or caruncle and/or tumor of the bulbar conjunctiva, more than 2 mm in thickness
[ ]	[ ]	pT4	Tumor invades eyelid, cornea, and/or orbit
		Lymph Node (pN)	
[ ]	[ ]	pNX	Regional lymph nodes cannot be assessed
[ ]	[ ]	pN0	No regional lymph node metastasis
[ ]	[ ]	pN1	Regional lymph node metastasis
		Distant Metastasis (pM)	
[ ]	[ ]	pMX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	pM0	No distant metastasis
[ ]	[ ]	pM1	Distant metastasis

Stage Grouping

No stage grouping is presently recommended.

Staged by _____ M.D.
_____ Registrar
Date _____

(continued on next page)

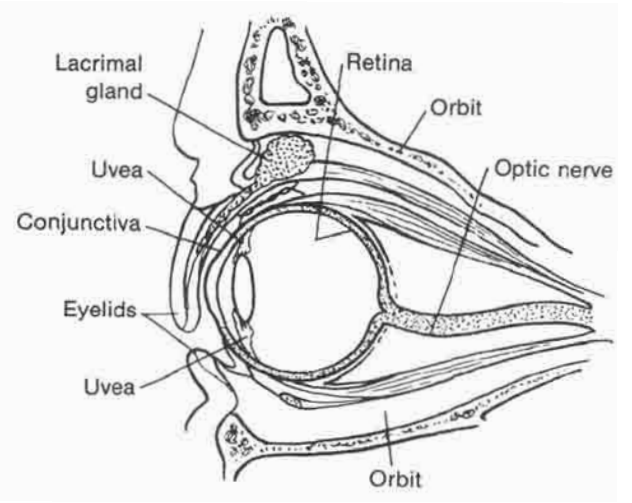
Histopathologic Type

The categorization applies only to malignant melanoma of the conjunctiva.

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G0 Primary acquired melanosis
[ ] G1 Malignant melanoma arising from a nevus
[ ] G2 Malignant melanoma arising from primary acquired melanosis
[ ] G3 Malignant melanoma arising de novo

Illustration



Indicate on diagram and describe exact location and characteristics of tumor.

42

Malignant Melanoma of the Uvea

C69.3 Choroid
C69.4 Ciliary body and iris

The classification applies only to melanoma.

ANATOMY

Primary Site. The middle layer of the eyeball, the uvea (uveal tract) lies between the cornea and sclera externally and the retina and its analogues internally. The uveal tract is divided into three regions: iris, ciliary body, and choroid. It is a highly vascular structure, with the choroid in particular composed of large blood vessels with little intervening connective tissue. There are no lymphatic channels in the uvea. Systemic metastasis from uveal melanomas occurs by hematogenous routes. Uveal melanomas are believed to arise from uveal melanocytes and are, therefore, of neural crest origin. Melanomas may spread by local extension through Bruch's membrane to involve the retina and vitreous, or by extension through the sclera or optic nerve into the orbit.

Most uveal melanomas occur in the choroid. The ciliary body is less commonly the site of origin, and the iris is least commonly involved. Iris melanomas are relatively benign and slow growing, and they rarely metastasize. Melanomas of the ciliary body and choroid are cytologically more malignant and metastasize more frequently.

It may be clinically impossible to distinguish a large nevus from a small melanoma.

Regional Lymph Nodes. Because there are no intraocular lymphatics, this category applies only to extrascleral extension anteriorly. The regional lymph nodes are:

Parotid
Preauricular
Submandibular
Cervical

Nodal involvement implies subconjunctival extension of the primary tumor.

Metastatic Sites. Uveal melanomas can metastasize through hematogenous routes to various organs. The liver is most commonly involved and usually is the first site of clinically detectable metastasis. Less commonly, the lung, pleura, subcutaneous tissues, bone, and other sites may be involved.

RULES FOR CLASSIFICATION

There should be histologic verification of the disease. Any unconfirmed case must be reported separately.

Clinical Staging. The assessment of the tumor is based on clinical examination, including slit-lamp examination and direct and indirect ophthalmoscopy. Additional methods such as ultrasonography, computerized stereometry, fluorescein angiography, and isotope examination may enhance the accuracy of appraisal.

Pathologic Staging. Complete resection of the primary site is indicated. Histologic study of the margins and the deep aspect of resected tissues is necessary. Resection or needle biopsy of enlarged regional lymph nodes or orbital masses is desirable.

DEFINITION OF TNM

These definitions apply to both clinical and pathologic staging.

ANATOMIC SITES

Iris
Ciliary body
Choroid

Iris

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
T1 Tumor limited to the iris
T2 Tumor involves one quadrant or less, with invasion into the anterior chamber angle
T3 Tumor involves more than one quadrant, with invasion into the anterior chamber angle
T4 Tumor with extraocular extension

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

Ciliary Body

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
T1 Tumor limited to the ciliary body
T2 Tumor invades into the anterior chamber and/or iris
T3 Tumor invades the choroid
T4 Tumor with extraocular extension

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

Choroid

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
T1* Tumor 10 mm or less in greatest dimension, with an elevation of 3 mm or less
T1a Tumor 7 mm or less in greatest dimension, with an elevation of 2 mm or less
T1b Tumor more than 7 mm but not more than 10 mm in greatest dimension, with an elevation of more than 2 mm but not more than 3 mm
T2* Tumor more than 10 mm but not more than 15 mm in greatest dimension, with an elevation of more than 3 mm but not more than 5 mm
T3* Tumor more than 15 mm in greatest dimension or with an elevation of more than 5 mm
T4 Tumor with extraocular extension

Note: When dimension and elevation show a difference in classification, the highest category should be used for classification.

* *Note:* In clinical practice, the tumor base may be estimated in optic disc diameters (dd) (average: 1 dd = 1.5 mm). The elevation may be estimated in diopters (average: 3 diopters = 1 mm). Other techniques, such as ultrasonography and computerized stereometry, may provide a more accurate measurement.

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

STAGE GROUPING

The classification of the structure most affected is used when more than one of the uveal structures is involved by tumor.

Iris and Ciliary Body

Stage I	T1	N0	M0
Stage II	T2	N0	M0
Stage III	T3	N0	M0
Stage IVA	T4	N0	M0
Stage IVB	Any T	N1	M0
	Any T	Any N	M1

Choroid

Stage IA	T1a	N0	M0
Stage IB	T1b	N0	M0
Stage II	T2	N0	M0
Stage III	T3	N0	M0
Stage IVA	T4	N0	M0
Stage IVB	Any T	N1	M0
	Any T	Any N	M1

HISTOPATHOLOGIC TYPE

The histopathologic types are:

Spindle cell melanoma
Mixed cell melanoma
Epithelioid cell melanoma

HISTOPATHOLOGIC GRADE (G)

GX Grade cannot be assessed
G1 Spindle cell melanoma
G2 Mixed cell melanoma
G3 Epithelioid cell melanoma

Venous Invasion (V)

VX Venous invasion cannot be assessed
V0 Veins do not contain tumor
V1 Veins in melanoma contain tumor
V2 Vortex veins contain tumor

Scleral Invasion (S)

SX Scleral invasion cannot be assessed
S0 Sclera does not contain tumor
S1 Intrascleral* invasion of tumor
S2 Extrascleral extension of tumor

**Note:* Includes perineural and perivascular invasion of scleral canals.

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	DEFINITIONS
		Iris
		Primary Tumor (T)
[ ]	[ ]	TX Primary tumor cannot be assessed
[ ]	[ ]	T0 No evidence of primary tumor
[ ]	[ ]	T1 Tumor limited to the iris
[ ]	[ ]	T2 Tumor involves one quadrant or less, with invasion into the anterior chamber angle
[ ]	[ ]	T3 Tumor involves more than one quadrant, with invasion into the anterior chamber angle
[ ]	[ ]	T4 Tumor with extraocular extension
		Lymph Node (N)
[ ]	[ ]	NX Regional lymph nodes cannot be assessed
[ ]	[ ]	N0 No regional lymph node metastasis
[ ]	[ ]	N1 Regional lymph node metastasis
		Distant Metastasis (M)
[ ]	[ ]	MX Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0 No distant metastasis
[ ]	[ ]	M1 Distant metastasis
		Ciliary Body
		Primary Tumor (T)
[ ]	[ ]	TX Primary tumor cannot be assessed
[ ]	[ ]	T0 No evidence of primary tumor
[ ]	[ ]	T1 Tumor limited to ciliary body
[ ]	[ ]	T2 Tumor invades into anterior chamber and/or iris
[ ]	[ ]	T3 Tumor invades choroid
[ ]	[ ]	T4 Tumor with extraocular extension
		Lymph Node (N)
[ ]	[ ]	NX Regional lymph nodes cannot be assessed
[ ]	[ ]	N0 No regional lymph node metastasis
[ ]	[ ]	N1 Regional lymph node metastasis
		Distant Metastasis (M)
[ ]	[ ]	MX Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0 No distant metastasis
[ ]	[ ]	M1 Distant metastasis
		Choroid
		Primary Tumor (T)
[ ]	[ ]	TX Primary tumor cannot be assessed
[ ]	[ ]	T0 No evidence of primary tumor
[ ]	[ ]	T1 Tumor 10 mm or less in greatest dimension with an elevation 3 mm or less
[ ]	[ ]	T1a Tumor 7 mm or less in greatest dimension with an elevation 2 mm or less
[ ]	[ ]	T1b Tumor more than 7 mm but not more than 10 mm in greatest dimension with an elevation more than 2 mm but not more than 3 mm
[ ]	[ ]	T2 Tumor more than 10 mm but not more than 15 mm in greatest dimension with an elevation of more than 3 mm but not more than 5 mm
[ ]	[ ]	T3 Tumor more than 15 mm in greatest dimension or with an elevation more than 5 mm
[ ]	[ ]	T4 Tumor with extraocular extension

(continued on next page)

Clin	Path
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]

Lymph Node (N)

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

Distant Metastasis (M)

MX	Presence of distant metastasis cannot be assessed
M0	No distant metastasis
M1	Distant metastasis

Clin	Path
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]
[ ]	[ ]

Stage Grouping

Iris and Ciliary Body

I	T1	N0	M0
II	T2	N0	M0
III	T3	N0	M0
IVA	T4	N0	M0
IVB	Any T	N1	M0
	Any T	Any N	M1

Choroid

IA	T1a	N0	M0
IB	T1b	N0	M0
II	T2	N0	M0
III	T3	N0	M0
IVA	T4	N0	M0
IVB	Any T	N1	M0
	Any T	Any N	M1

Staged by _____ M.D.

_____ Registrar

Date _____

Histopathologic Type

Spindle cell melanoma
Mixed cell melanoma
Epithelioid cell melanoma

Histopathologic Grade (G)

[ ]	GX	Grade cannot be assessed
[ ]	G1	Spindle cell melanoma
[ ]	G2	Mixed cell melanoma
[ ]	G3	Epithelioid cell melanoma

Venous Invasion (V)

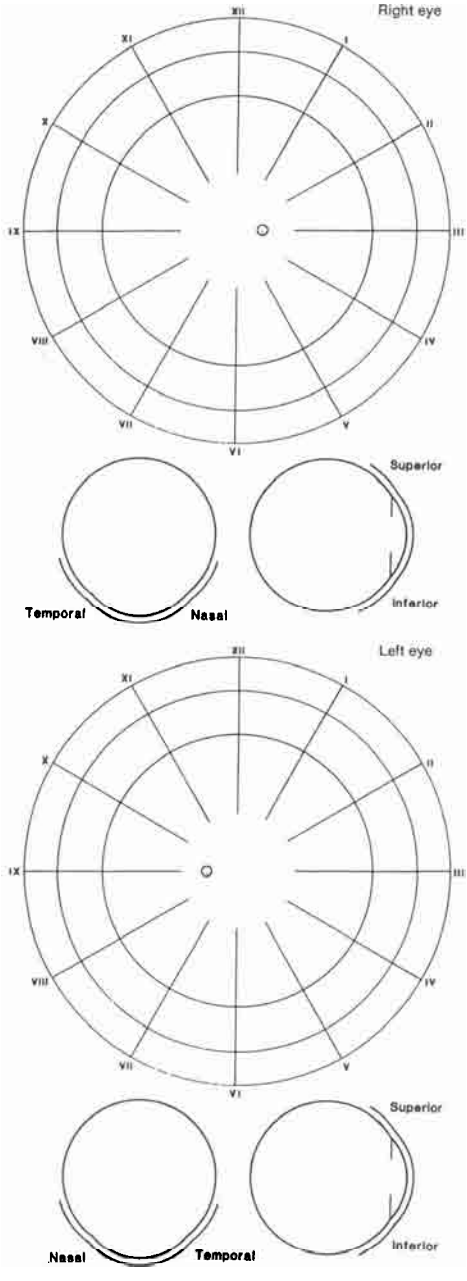
[ ]	VX	Venous invasion cannot be assessed
[ ]	V0	Veins do not contain tumor
[ ]	V1	Veins in melanoma contain tumor
[ ]	V2	Vortex veins contain tumor

Scleral Invasion (S)

[ ]	SX	Scleral invasion cannot be assessed
[ ]	S0	Sclera does not contain tumor
[ ]	S1	Intrascleral invasion of tumor
[ ]	S2	Extrascleral invasion of tumor*

* *Note:* Includes perineural and perivascular invasion of scleral canals.

Illustrations



Indicate on diagrams and describe exact location and characteristics of tumor.

43

Retinoblastoma

C69.2 Retina

ANATOMY

Primary Site. The retina comprises neurons and glial cells. The neurons give rise to retinoblastoma, whereas the glial cells give rise to astrocytomas, which in the retina are benign and extremely rare. The retina is limited internally by a membrane separating it from the vitreous cavity. Externally, it is limited by the retinal pigment epithelium and Bruch's membrane, which separate it from the choroid and act as natural barriers to extension of retinal tumors into the choroid. The continuation of the retina with the optic nerve allows direct extension of retinoblastomas into the optic nerve and then to the subarachnoid space. Because the retina has no lymphatics, spread of retinal tumors occurs either by direct extension into adjacent structures or by distant metastasis through hematogenous routes.

Regional Lymph Nodes. Because there are no intraocular lymphatics, the category applies only to anterior extrascleral extension. The regional lymph nodes are:

Parotid
Preauricular
Submandibular
Cervical

Involvement implies subconjunctival extension of the tumor.

Metastatic Sites. Retinoblastoma can metastasize through hematogenous routes to various sites, most notably the skull, long bones, brain, lymph nodes, and viscera.

RULES FOR CLASSIFICATION

Clinical Staging. In bilateral cases, each eye must be classified separately. The classification does not apply to complete spontaneous regression of the tumor. There should be histologic verification of the disease in an enucleated eye. Any unconfirmed case must be reported separately. The extent of retinal involvement is indicated as a percentage.

Pathologic Staging. All clinical and pathologic data from the resected specimen are to be used.

DEFINITION OF TNM

Clinical Classification (cTNM)

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
T1 Tumor(s) limited to 25% or less of the retina
T2 Tumor(s) involve(s) more than 25% but not more than 50% of the retina
T3 Tumor(s) involve(s) more than 50% of the retina and/or invade(s) beyond the retina but remain(s) intraocular
- T3a Tumor(s) involve(s) more than 50% of the retina and/ or tumor cells in the vitreous
- T3b Tumor(s) involve(s) the optic disc
- T3c Tumor(s) involve(s) the anterior chamber and/or uvea

T4 Tumor with extraocular invasion
T4a Tumor invades the retrobulbar optic nerve
T4b Extraocular extension other than invasion of the optic nerve

Note: The following suffixes may be added to the appropriate T categories: "m" indicates multiple tumors (e.g., T2 [m2]); "f" indicates cases with a known family history; and "d" indicates diffuse retinal involvement without the formation of discrete masses.

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

Pathologic Classification (pTNM)

Primary Tumor (pT)

pTX Primary tumor cannot be assessed
pT0 No evidence of primary tumor
pT1 Tumor(s) limited to 25% or less of the retina
pT2 Tumor(s) involve(s) more than 25% but not more than 50% of the retina
pT3 Tumor(s) involve(s) more than 50% of the retina and/or invade(s) beyond the retina but remain(s) intraocular
- pT3a Tumor(s) involve(s) more than 50% of the retina and/ or tumor cells in the vitreous
- pT3b Tumor invades the optic nerve as far as the lamina cribrosa
- pT3c Tumor in the anterior chamber and/or invasion with thickening of the uvea and/or intrascleral invasion

pT4 Tumor with extraocular invasion
- pT4a Intraneural tumor beyond the lamina cribrosa but not at the line of resection
- pT4b Tumor at the line of resection or other extraocular extension

Regional Lymph Nodes (pN)

pNX Regional lymph nodes cannot be assessed
pN0 No regional lymph node metastasis
pN1 Regional lymph node metastasis

Distant Metastasis (pM)

pMX Presence of distant metastasis cannot be assessed
pM0 No distant metastasis
pM1 Distant metastasis

STAGE GROUPING

In cases of bilateral disease, the more affected eye is used for the stage grouping.

Stage IA	T1	N0	M0
Stage IB	T2	N0	M0
Stage IIA	T3a	N0	M0
Stage IIB	T3b	N0	M0
Stage IIC	T3c	N0	M0
Stage IIIA	T4a	N0	M0
Stage IIIB	T4b	N0	M0
Stage IV	Any T	N1	M0
	Any T	Any N	M1

Note: Pathologic stage grouping corresponds to the clinical stage grouping.

HISTOPATHOLOGIC TYPE

This classification applies only to retinoblastoma.

Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	DEFINITIONS
		Primary Tumor (T)
[ ]	[ ]	TX Primary tumor cannot be assessed
[ ]	[ ]	T0 No evidence of primary tumor
[ ]	[ ]	T1 Tumor(s) limited to 25% or less of the retina
[ ]	[ ]	T2 Tumor(s) involve(s) more than 25% but not more than 50% of the retina
[ ]	[ ]	T3 Tumor(s) involve(s) more than 50% of the retina and/or invade(s) beyond the retina but remain(s) intraocular
[ ]	[ ]	T3a Tumor(s) involve(s) more than 50% of the retina and/or tumor cells in the vitreous
[ ]	[ ]	T3b Tumor(s) involve(s) optic disc
[ ]	[ ]	T3c Tumor(s) involve(s) anterior chamber and/or uvea
[ ]	[ ]	T4 Tumor with extraocular invasion
[ ]	[ ]	T4a Tumor invades retrobulbar optic nerve
[ ]	[ ]	T4b Extraocular extension other than invasion of optic nerve
		Lymph Node (N)
[ ]	[ ]	NX Regional lymph nodes cannot be assessed
[ ]	[ ]	N0 No regional lymph node metastasis
[ ]	[ ]	N1 Regional lymph node metastasis
		Distant Metastasis (M)
[ ]	[ ]	MX Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0 No distant metastasis
[ ]	[ ]	M1 Distant metastasis
		Primary Tumor (pT)
[ ]	[ ]	pTX Primary tumor cannot be assessed
[ ]	[ ]	pT0 No evidence of primary tumor
[ ]	[ ]	pT1 Tumor(s) limited to 25% or less of the retina
[ ]	[ ]	pT2 Tumor(s) involve(s) more than 25% but not more than 50% of the retina
[ ]	[ ]	pT3 Tumor(s) involve(s) more than 50% of the retina and/or invade(s) beyond the retina but remain(s) intraocular
[ ]	[ ]	pT3a Tumor(s) involve(s) more than 50% of the retina and/or tumor cells in the vitreous
[ ]	[ ]	pT3b Tumor invades optic nerve as far as the lamina cribrosa
[ ]	[ ]	pT3c Tumor in anterior chamber and/or invasion with thickening of the uvea and/or intrascleral invasion
[ ]	[ ]	pT4 Tumor with extraocular invasion
[ ]	[ ]	pT4a Intraneural tumor beyond the lamina cribrosa but not at the line of resection
[ ]	[ ]	pT4b Tumor at the line of resection or other extraocular extension
		Lymph Node (pN)
[ ]	[ ]	pNX Regional lymph nodes cannot be assessed
[ ]	[ ]	pN0 No regional lymph node metastasis
[ ]	[ ]	pN1 Regional lymph node metastasis
		Distant Metastasis (pM)
[ ]	[ ]	pMX Presence of distant metastasis cannot be assessed
[ ]	[ ]	pM0 No distant metastasis
[ ]	[ ]	pM1 Distant metastasis

(continued on next page)

Clin	Path	Stage Grouping			
[ ]	[ ]	IA	T1	N0	M0
[ ]	[ ]	IB	T2	N0	M0
[ ]	[ ]	IIA	T3a	N0	M0
[ ]	[ ]	IIB	T3b	N0	M0
[ ]	[ ]	IIC	T3c	N0	M0
[ ]	[ ]	IIIA	T4a	N0	M0
[ ]	[ ]	IIIB	T4b	N0	M0
[ ]	[ ]	IV	Any T	N1	M0
			Any T	Any N	M1

Staged by _____ M.D.

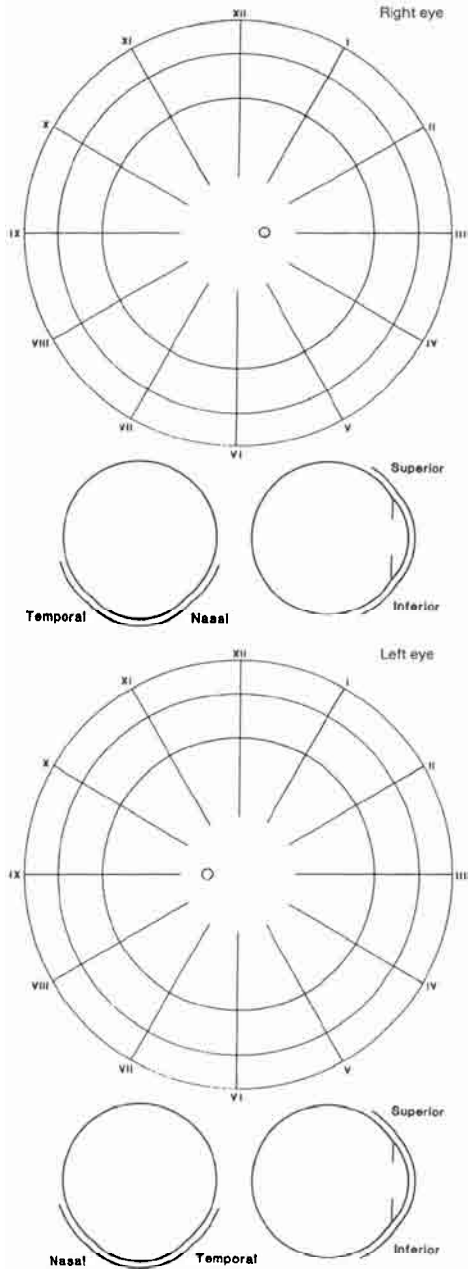
_____ Registrar

Date _____

Histopathologic Type

This classification applies only to retinoblastoma.

Illustrations



Indicate on diagrams and describe exact location and characteristics of tumor.

_____

_____

_____

_____

44

Sarcoma of the Orbit

C69.6 Orbit, NOS
C69.8 Overlapping lesion

Sarcomas of the orbit include a broad spectrum of soft-tissue tumors and sarcomas of bone.

ANATOMY

Primary Site. Sarcoma of the orbit occurs in the soft tissues and bone of the orbital fossa.

Regional Lymph Nodes. The regional lymph nodes are:

Submandibular
Parotid (preauricular)
Cervical

Metastatic Sites. Metastatic spread occurs by way of the bloodstream to distant sites.

RULES FOR CLASSIFICATION

Clinical Staging. Clinical classification is based on symptoms and signs relating to visual loss, degree of proptosis or displacement, papilledema, and optic atrophy. Diagnostic tests include radiographs of the orbit, computed tomography, and angiography.

Pathologic Staging. Pathologic classification is based on the histopathology of the tumor, its grade, and the extent of removal.

DEFINITION OF TNM

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
T1 Tumor 15 mm or less in greatest dimension
T2 Tumor more than 15 mm in greatest dimension
T3 Tumor of any size with diffuse invasion of orbital tissues and/or bony walls
T4 Tumor invades beyond the orbit to adjacent sinuses and/ or to the cranium

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

STAGE GROUPING

No stage grouping is presently recommended.

HISTOPATHOLOGIC TYPE

Sarcomas of the orbit include a broad spectrum of soft-tissue tumors and sarcomas of bone.

HISTOPATHOLOGIC GRADE (G)

GX Grade cannot be assessed
G1 Well differentiated
G2 Moderately differentiated
G3 Poorly differentiated
G4 Undifferentiated

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Data Form for Cancer Staging

Patient identification
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Address _____
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Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

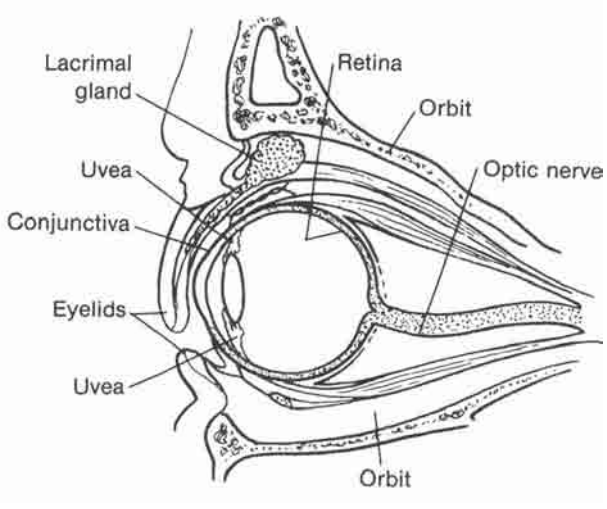
Clin	Path	DEFINITIONS	
		Primary Tumor (T)	
[ ]	[ ]	TX	Primary tumor cannot be assessed
[ ]	[ ]	T0	No evidence of primary tumor
[ ]	[ ]	T1	Tumor 15 mm or less in greatest dimension
[ ]	[ ]	T2	Tumor more than 15 mm in greatest dimension
[ ]	[ ]	T3	Tumor of any size with diffuse invasion of orbital tissues and/or bony walls
[ ]	[ ]	T4	Tumor invades beyond the orbit to adjacent sinuses and/or to cranium
		Lymph Node (N)	
[ ]	[ ]	NX	Regional lymph nodes cannot be assessed
[ ]	[ ]	N0	No regional lymph node metastasis
[ ]	[ ]	N1	Regional lymph node metastasis
		Distant Metastasis (M)	
[ ]	[ ]	MX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0	No distant metastasis
[ ]	[ ]	M1	Distant metastasis

Stage Grouping

No stage grouping is presently recommended

Staged by _____ M.D.
_____ Registrar
Date _____

Illustration



Indicate on diagrams and describe exact location and characteristics of tumor.

_____
_____
_____
_____

Histopathologic Type

Sarcomas of the orbit include a broad spectrum of soft-tissue tumors and sarcomas of bone.

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated
[ ] G3 Poorly differentiated
[ ] G4 Undifferentiated

45

Carcinoma of the Lacrimal Gland

C69.5 Lacrimal gland

A retrospective study of 265 epithelial tumors of the lacrimal gland has been completed from material on file in the Registry of Ophthalmic Pathology at the Armed Forces Institute of Pathology. The histologic classification used is a modification of the WHO classification of salivary gland tumors. The lacrimal gland includes both lobules: the superficial (palpebral lobe) portion and the deep intraorbital portion.

ANATOMY

Primary Site. The lacrimal gland lies in a bony excavation covered by periosteum, located in the lateral orbital wall (the fossa of the lacrimal gland). The smaller palpebral portion projects into the lateral portion of the upper lid between the palpebral fascia and the conjunctiva.

Regional Lymph Nodes. The regional lymph nodes include:

Parotid (preauricular)
Submandibular
Cervical

Metastatic Sites. The lung is the most common metastatic site, followed by bone and remote viscera.

RULES FOR CLASSIFICATION

Clinical Staging. A complete physical examination, imaging of the orbit (including computed tomography, ultrasonography, and plane films), and tomography of the adjacent paranasal sinuses should be done. Chest x-ray films, radionuclide bone scans, and blood chemistries should also be available.

Pathologic Staging. After complete resection of the mass, the entire specimen should be evaluated to determine the type of tumor and the grade of malignancy.

DEFINITION OF TNM

This classification applies to both clinical and pathologic staging.

Primary Tumor (T)

- TX Primary tumor cannot be assessed
- T0 No evidence of primary tumor
- T1 Tumor 2.5 cm or less in greatest dimension limited to the lacrimal gland
- T2 Tumor 2.5 cm or less in greatest dimension invading the periosteum of the fossa of the lacrimal gland
- T3 Tumor more than 2.5 cm but not more than 5 cm in greatest dimension
  - T3a Tumor limited to the lacrimal gland
  - T3b Tumor invades the periosteum of the fossa of the lacrimal gland
- T4 Tumor more than 5 cm in greatest dimension
  - T4a Tumor invades the orbital soft tissues, optic nerve, or globe without bone invasion
  - T4b Tumor invades the orbital soft tissues, optic nerve, or globe with bone invasion

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

- MX Presence of distant metastasis cannot be assessed
- M0 No distant metastasis
- M1 Distant metastasis

STAGE GROUPING

No stage grouping is presently recommended.

HISTOPATHOLOGIC TYPE

The major malignant primary epithelial tumors include the following:

Carcinoma in pleomorphic adenoma (malignant mixed tumor), which includes adenocarcinoma and adenoid cystic carcinoma arising in benign mixed tumor (BMT)
Adenoid cystic carcinoma (cylindroma), arising de novo
Adenocarcinoma, arising de novo
Mucoepidermoid carcinoma
Squamous cell carcinoma

HISTOPATHOLOGIC GRADE (G)

GX Grade cannot be assessed
G1 Well differentiated
G2 Moderately differentiated: includes adenoid cystic carcinoma without baseloid (solid) pattern
G3 Poorly differentiated: includes adenoid cystic carcinoma with baseloid (solid) pattern
G4 Undifferentiated

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Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	DEFINITIONS
		Primary Tumor (T)
[ ]	[ ]	TX Primary tumor cannot be assessed
[ ]	[ ]	T0 No evidence of primary tumor
[ ]	[ ]	T1 Tumor 2.5 cm or less in greatest dimension limited to the lacrimal gland and mobile within the lacrimal fossa
[ ]	[ ]	T2 Tumor 2.5 cm or less in greatest dimension invading the periosteum of the fossa of the lacrimal gland
[ ]	[ ]	T3 Tumor more than 2.5 cm but not more than 5 cm in greatest dimension
[ ]	[ ]	T3a Tumor limited to the lacrimal gland
[ ]	[ ]	T3b Tumor invades the periosteum of the fossa of the lacrimal gland
[ ]	[ ]	T4 Tumor more than 5 cm in greatest dimension
[ ]	[ ]	T4a With invasion of orbital soft tissues, optic nerve, or globe, *without* bone invasion
[ ]	[ ]	T4b With invasion of orbital soft tissues, optic nerve, or globe, *with* bone invasion
		Lymph Node (N)
[ ]	[ ]	NX Regional lymph nodes cannot be assessed
[ ]	[ ]	N0 No regional lymph node metastasis
[ ]	[ ]	N1 Regional lymph node metastasis
		Distant Metastasis (M)
[ ]	[ ]	MX Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0 No distant metastasis
[ ]	[ ]	M1 Distant metastasis

Stage Grouping

No stage grouping is presently recommended.

Staged by _____ M.D.
_____ Registrar
Date _____

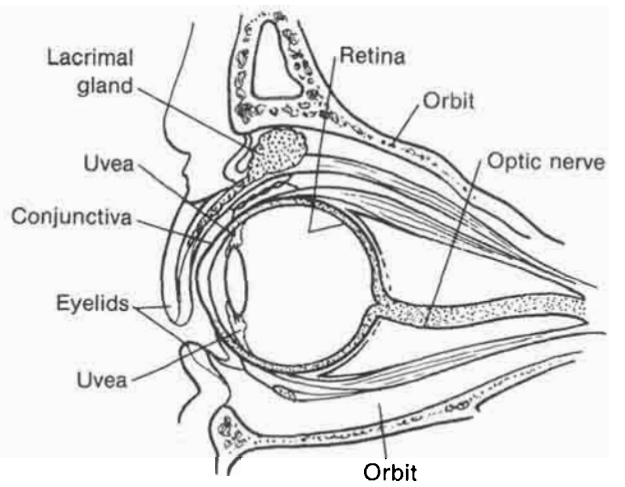
Histopathologic Type

The major malignant primary epithelial tumors include the following:
Carcinoma in pleomorphic adenoma (malignant mixed tumor), which includes adenocarcinoma and adenoid cystic carcinoma arising in benign mixed tumor (BMT)
Adenoid cystic carcinoma (cylindroma) arising de novo
Adenocarcinoma (arising de novo)
Mucoepidermoid carcinoma
Squamous cell carcinoma

Histopathologic Grade (G)

[ ] GX Grade cannot be assessed
[ ] G1 Well differentiated
[ ] G2 Moderately differentiated: includes adenoid cystic carcinoma without baseloid (solid) pattern
[ ] G3 Poorly differentiated: includes adenoid cystic carcinoma with baseloid (solid) pattern
[ ] G4 Undifferentiated

Illustration



Indicate on diagram and describe exact location and characteristics of tumor.

CENTRAL NERVOUS SYSTEMS

46

Brain

C70.0 Cerebral meninges

C71.0 Cerebrum
C71.1 Frontal lobe
C71.2 Temporal lobe
C71.3 Parietal lobe
C71.4 Occipital lobe
C71.5 Ventricle, NOS
C71.6 Cerebellum, NOS
C71.7 Brain stem
C71.8 Overlapping lesion
C71.9 Brain, NOS

The most critical feature in the classification of brain tumors is histopathology. Accurate pathologic criteria and classification are essential to an understanding of the clinical and biologic behavior of the gliomas in particular, and of most other tumors as well. The anatomic location and extent of tumors within the brain are also of clinical and prognostic significance. Neuroradiologic-diagnostic procedures have become increasingly more accurate and reliable in providing topographic and morphologic information on tumors of the brain and are useful at various points in diagnosis and management. The recommendations in this chapter refer to primary tumors of the brain. A system for staging metastatic tumors of the brain is under development and is currently being tested.

ANATOMY

Primary Site. Various tissues within the brain can give rise to neoplasms, including astrocytes and other glial cells, meninges, blood vessels, pituitary and pineal cells, and neural elements proper. The major structural sites involved include the various lobes of the cerebral hemispheres; the midline structures, including the midbrain, pons, and medulla; and the posterior fossa.

Regional Lymph Nodes. There are no lymphatic structures draining the brain.

Metastatic Sites. Certain brain tumors can seed into the subarachnoid space. Hematogenous spread is very uncommon but on rare occasions has occurred in bone and other sites.

RULES FOR CLASSIFICATION

Clinical Staging. This staging is based on neurologic signs and symptoms and on neurologic diagnostic tests, including skull radiography, electroencephalography, isotopic brain scans, cerebral angiography, pneumoencephalography, computed tomography, and magnetic resonant imaging. All diagnostic information available prior to first definitive treatment may be used.

Pathologic Staging. This staging is based on histopathology, grade, and microscopic evidence of completeness of resected tumor removal.

DEFINITION OF TNM

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor

Supratentorial Tumor

T1 Tumor 5 cm or less in greatest dimension; limited to one side

T2 Tumor more than 5 cm in greatest dimension; limited to one side
T3 Tumor invades or encroaches on the ventricular system
T4 Tumor crosses the midline, invades the opposite hemisphere, or invades infratentorially

Infratentorial Tumor

T1 Tumor 3 cm or less in greatest dimension; limited to one side
T2 Tumor more than 3 cm in greatest dimension; limited to one side
T3 Tumor invades or encroaches on the ventricular system
T4 Tumor crosses the midline, invades the opposite hemisphere, or invades supratentorially

Regional Lymph Nodes (N)

This category does not apply to this site.

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

HISTOPATHOLOGIC GRADE (G)

GX Grade cannot be assessed
G1 Well differentiated
G2 Moderately differentiated
G3 Poorly differentiated
G4 Undifferentiated

STAGE GROUPING

Stage IA	G1	T1	M0
Stage IB	G1	T2	M0
	G1	T3	M0
Stage IIA	G2	T1	M0
Stage IIB	G2	T2	M0
	G2	T3	M0
Stage IIIA	G3	T1	M0
Stage IIIB	G3	T2	M0
	G3	T3	M0
Stage IV	G1	T4	M0
	G2	T4	M0
	G3	T4	M0
	G4	Any T	M0
	Any G	Any T	M1

HISTOPATHOLOGIC TYPE

Tumors included in analysis and evaluation are:

Astrocytomas
Oligodendrogliomas
Ependymal and choroid plexus tumors
Glioblastomas
Medulloblastomas
Meningiomas, malignant
Neurilemmomas (neurinomas, schwannomas), malignant
Hemangioblastomas
Neurosarcomas
Other sarcomas

Histologic grade usually correlates with biologic activity of the tumor. This is particularly the case with malignant astrocytomas, the most common form of glioma. The age of the patient at the time of diagnosis is also of major importance for prognosis.

APPENDIX

Histologic Grading of Tumors of the Central Nervous System

Criteria for the Diagnosis of Malignancy in Tumors of the Central Nervous System and Allied Structures

For tumors of the central nervous system and allied structures, the uncritical application of criteria for histologic and biologic malignancy that generally pertain to other neoplasms is inadequate for the following reasons:

1. Irrespective of the histologic malignancy of the tumor, its unimpeded growth within the confines of the skull as a space-occupying and expanding lesion inevitably leads to a fatal termination, which by definition is equated with clinical malignancy.
2. Similarly, the local pressure caused by an intracranial tumor on vital neural structures may result in the clinical effects of malignancy, irrespective of the histologic type of tumor.
3. The obstructive effect of a growing tumor leads to the production of secondary occlusive hydrocephalus.
4. Certain criteria of malignancy of neoplasms that in other body systems pertain to their growth and spread (especially the characteristic of infiltrative growth and the capacity to metastasize, either within or outside the central nervous system) do not necessarily pertain to, or have to be modified to, the evaluation of the malignant behavior of central nervous system tumors.

Thus, tumors of the central nervous system and allied structures, in addition to their intrinsic benign or malignant histologic character that to a considerable extent

determines their biologic behavior, may by their specific localization acquire certain characteristics that collectively will add up to a picture regarded as benign, semibenign, relatively malignant, or highly malignant.

The numerical grading used in this classification is based on histologic criteria of malignancy and should be considered an estimate of the usual behavior of each type of tumor. Numerical grade 1 is considered the least malignant; grades 2, 3, and 4 indicate increasing degrees of malignancy.

In this general evaluation, the pathologist confronted with the problem of malignancy and prognosis is faced with two sets of data. In the first analysis, the evaluation of malignancy must clearly be based on retrospective assessment of the postoperative prognosis and survival rates of other known similar examples, leading to a final and reasonably accurate clinicopathologic correlation that both reinforces the purely histopathologic evaluation of malignancy and is reinforced by it.

Second, the pathologist deduces malignancy from a number of purely histologic and cytologic data. These include increase of cellularity, presence and rate of mitotic figures, presence of atypical mitotic figures, pleomorphism of tumor cells, pleomorphism of tissue architecture (particularly necroses, abnormally prominent stromal reaction, disorderly stromal reaction, and overgrowth), and the formation of pathologic blood vessels (corresponding to the angiographic appearance of arteriovenous fistulas).

On the other hand, other features usually regarded as indicative of or synonymous with malignancy need not necessarily be recognized in the case of tumors of the central nervous system, especially those of neuroectodermal origin. For instance, lack of circumscription and focal parenchymatous invasion is not a necessary accompaniment of cellular anaplasia or ultimate clinical malignancy. Also, the actual presence of mitotic figures (as in oligodendroglioma) does not necessarily imply a particularly malignant behavior; the overall number of mitoses and the presence of abnormal mitotic figures are more important in evaluation. Similarly, local invasion of the leptomeninges is often clearly dissociated from either of the two features just quoted. This is the case, for example, in the pilocytic astrocytoma that involves the wall of the third ventricle, the optic nerve, the cerebellum, and so on.

Although distant meningeal and ventricular metastases are often characteristic of highly malignant tumors such as medulloblastoma, this phenomenon again is not always to be correlated with the highest degrees of cytologic malignancy, as seen in some oligodendrogliomas.

The Question of Grading

Following Broder's classification of epithelial tumors elsewhere in the body, an attempt has been made by Kernohan and his school to apply a system of grading by ascending degrees of malignancy, numbered 1 to 4, to certain tumors of neuroectodermal origin—namely astrocytoma, oligodendroglioma, ependymoma, and neuroastrocytoma. This attempt stemmed both from a desire to simplify the then current classification of tumors of the central nervous system and from a need to offer to the neurosurgeon a prognostic evaluation of the tumor removed at surgery, based on certain definite histologic and cytologic criteria. Attractive though this attempt at simplification might be, however, it has to meet with a number of objections:

1. The sample of tissue so analyzed may from surgical necessity not be representative of the tumor as a whole.
2. The specific evolution of the particular tumor in terms of its anaplastic potentialities is not fully expressed by such a scheme of grading. For example, a cerebellar pilocytic astrocytoma graded 1 does not have the same anaplastic potential as a cerebral astrocytoma or some other tumors also graded 1.
3. The pleomorphism of cell and tissue structures so frequently inherent in primary neuroectodermal tumors poses additional difficulties to the application of a simplified system of grading.
4. This cytologic grading makes it extremely difficult to place tumors with mixed cell populations into an already predetermined tumor category.

Nevertheless, the above remarks should not be regarded as basically antagonistic to some attempts at expressing the degree of malignancy of a particular tumor of the central nervous system. Indeed, from the clinical and therapeutic points of view, no classification based on purely histologic entities is satisfactory unless adequate cognizance is taken of, and information provided on, the degree of malignancy of a particular tumor submitted for examination. Thus, it is the duty and prerogative of the pathologist to provide his clinical colleagues with an informed opinion on the likely evolution of a particular tumor, and to some extent this prognostic opinion is embodied in the recognition of specific clinicopathologic neuro-oncologic entities. As an illustration, it might be pointed out that two tumors of similar cellularity, isomorphous appearance, and mitotic rate—such as the medulloblastomas and some oligodendrogliomas—usually do not exhibit the same biologic behavior. This acquired body of knowledge is clearly the result of previous collaboration among clinicians and pathologists in the field of neuro-oncology.

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Data Form for Cancer Staging

Patient identification
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Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

DEFINITIONS

Primary Tumor (T)

Clin	Path		
[ ]	[ ]	TX	Primary tumor cannot be assessed
[ ]	[ ]	T0	No evidence of primary tumor

Supratentorial tumor

[ ]	[ ]	T1	Tumor 5 cm or less in greatest dimension; limited to one side
[ ]	[ ]	T2	Tumor more than 5 cm in greatest dimension; limited to one side
[ ]	[ ]	T3	Tumor invades or encroaches upon the ventricular system
[ ]	[ ]	T4	Tumor crosses the midline, invades the opposite hemisphere, or extends infratentorially

Infratentorial tumor

[ ]	[ ]	T1	Tumor 3 cm or less in greatest dimension; limited to one side
[ ]	[ ]	T2	Tumor more than 3 cm in greatest dimension; limited to one side
[ ]	[ ]	T3	Tumor invades or encroaches upon the ventricular system
[ ]	[ ]	T4	Tumor crosses the midline, invades the opposite hemisphere, or invades supratentorially

Lymph Node (N)

This category does not apply to this site.

Distant Metastasis (M)

[ ]	[ ]	MX	Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0	No distant metastasis
[ ]	[ ]	M1	Distant metastasis

Histopathologic Grade (G)

[ ]	[ ]	GX	Grade cannot be assessed
[ ]	[ ]	G1	Well differentiated
[ ]	[ ]	G2	Moderately differentiated
[ ]	[ ]	G3	Poorly differentiated
[ ]	[ ]	G4	Undifferentiated

Stage Grouping

Clin	Path				
]	[ ]	IA	G1	T1	M0
]	[ ]	IB	G1	T2	M0
			G1	T3	M0
]	[ ]	IIA	G2	T1	M0
]	[ ]	IIB	G2	T2	M0
			G2	T3	M0
]	[ ]	IIIA	G3	T1	M0
]	[ ]	IIIB	G3	T2	M0
			G3	T3	M0
]	[ ]	IV	G1	T4	M0
			G2	T4	M0
			G3	T4	M0
			G4	Any T	M0
			Any G	Any T	M1

Staged by _____ M.D.
_____ Registrar
Date _____

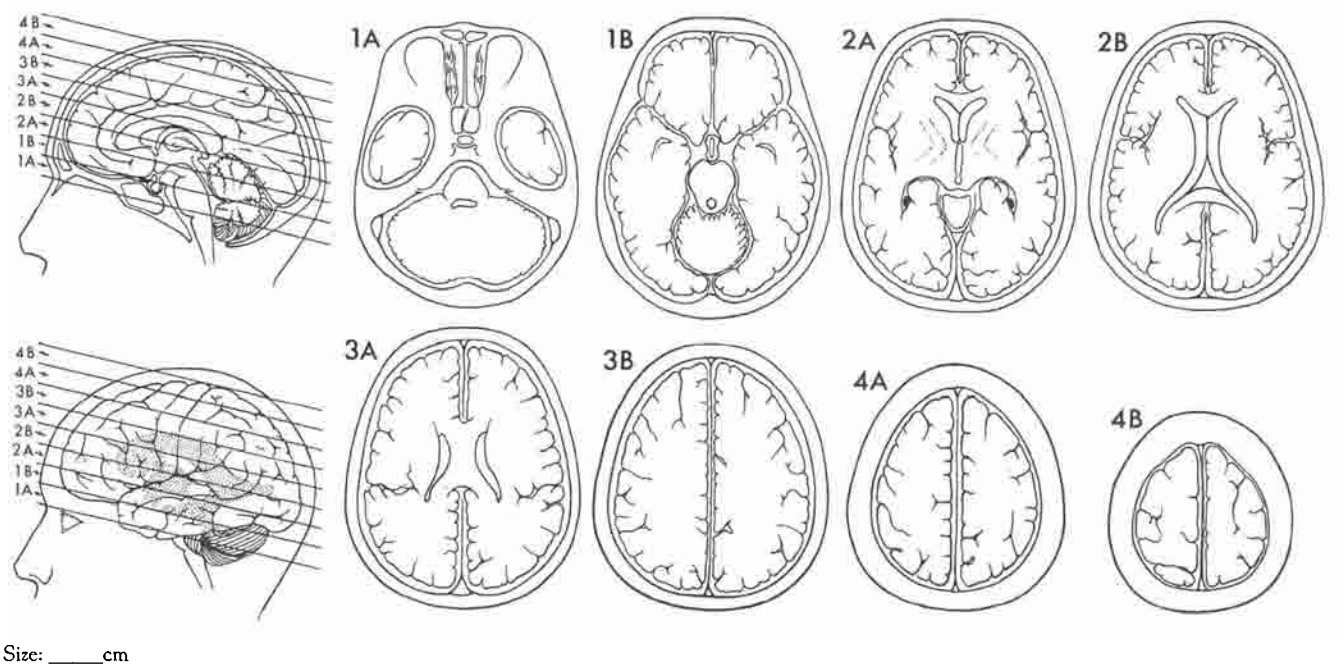
(continued on next page)

Histopathologic Type

Tumors that are included in the analysis and evaluation are as follows:

Astrocytoma
Oligodendroglioma
Ependymal and choroid plexus tumors
Glioblastoma
Medulloblastoma
Meningiomas, malignant
Neurilemmomas (neurinomas, schwannomas), malignant
Hemangioblastoma
Neurosarcomas
Other sarcomas

Illustrations



Size: _____cm
Weight: ——g

Indicate on diagrams the exact location and chasracteristics of tumor.

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Hodgkin's Disease

A distinctive form of lymphoma, Hodgkin's disease has served as a model for treatment trials, for great strides have been made in therapy for this disease. Staging of Hodgkin's lymphoma is not based on the local extent of disease but rather on its distribution and symptomatology. The classic TNM system is not useful for staging Hodgkin's disease. It is usually not possible to determine the primary tumor site. When the patient presents, the disease is often widely disseminated. Important for staging is the evaluation of many organs and groups of lymph nodes for tumor involvement. The disease is often associated with unusual immunologic abnormalities and a diversity of histologic changes. Staging is considered critical for patient management.

ANATOMY

The major lymphatic structures include groups and chains of lymph nodes, the spleen, and the thymus gland. The digestive system is also an important lymphoid organ that has collections of lymphoid tissue known as Waldeyer's ring in the oropharynx, Peyer's patches in the ileum, and lymphoid nodules in the appendix. Hodgkin's disease can involve almost any organ or tissue, especially the liver, bone marrow, and spleen, in addition to the lymph nodes.

RULES FOR CLASSIFICATION

Clinical Staging. The clinical stage is determined by obtaining an adequate initial biopsy, history, physical examination, laboratory tests, and imaging studies. Such studies usually establish the diagnosis and histologic type of Hodgkin's disease. Histologic confirmation is essential. All symptoms should be recorded, especially fever and weight loss.

Pathologic Staging. Pathologic staging depends on one or more lymph node biopsies, bone marrow biopsy, and, if the result will influence therapy, a laparotomy, which would include liver biopsy, splenectomy, and multiple nodal biopsies to assess distribution of the abdominal disease. Involved organs and sites should be listed.

STAGE GROUPING

Stage I	Involvement of single lymph node region (I) or localized involvement of a single extralymphatic organ or site (I_E).
Stage II	Involvement of two or more lymph node regions on the same side of the diaphragm (II) or localized involvement of a single associated extralymphatic organ or site and its regional lymph node(s) with or without involvement of other lymph node regions on the same side of the diaphragm (II_E).
	Note: The number of lymph node regions involved may be indicated by a subscript (e.g., II_3).
Stage III	Involvement of lymph node regions on both sides of the diaphragm (III), which may also be accompanied by localized involvement of an associated extralymphatic organ or site (III_E), by involvement of the spleen (III_S), or both $III_{(E+S)}$.
Stage IV	Disseminated (multifocal) involvement of one or more extralymphatic organs, with or without associated lymph node involvement, or isolated extralymphatic organ involvement with distant (nonregional) nodal involvement.

SYSTEMIC SYMPTOMS

Each stage is subdivided into "A" and "B" categories, "B" for those with defined systemic symptoms and "A" for those without. The B designation is given to those patients with (1) unexplained loss of more than 10% of body weight in the 6 months before diagnosis; (2) unexplained fever with temperatures above 38 C; and (3) drenching night sweats. Pruritus alone does not qualify

for B classification, nor does a short febrile illness associated with an infection.*

* *Note:* Pruritus as a systemic symptom remains controversial. This symptom is hard to define quantitatively and uniformly, but when it is recurrent, generalized, and otherwise unexplained, and when it ebbs and flows parallel to disease activity, it may be the equivalent of a B symptom.

HISTOPATHOLOGIC TYPE

Hodgkin's disease is divided into four major histologic types and "unclassified." These types should be recorded because they have prognostic significance. They are:

Nodular sclerosis
Lymphocyte predominance
Mixed cellularity
Lymphocyte depletion
Unclassified

Histologic classification should be based on paraffin-embedded hematoxylin and eosin-stained sections.

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Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	DEFINITIONS
		Stage Grouping
[ ]	[ ]	Stage I Involvement of single lymph node region (I) or localized involvement of a single extralymphatic organ or site (I_E).
[ ]	[ ]	Stage II Involvement of two or more lymph node regions on the same side of the diaphragm (II) or localized involvement of a single associated extralymphatic organ or site and its regional lymph node(s) with or without involvement of other lymph node regions on the same side of the diaphragm (II_E). NOTE: The number of lymph node regions involved may be indicated by a subscript (*e.g.*, II_3).
[ ]	[ ]	Stage III Involvement of lymph node regions on both sides of the diaphragm (III), which may also be accompanied by localized involvement of an associated extralymphatic organ or site (III_E), by involvement of the spleen (III_S), or both (III_{E+S}).
[ ]	[ ]	Stage IV Disseminated (multifocal) involvement of one or more extralymphatic organs, with or without associated lymph node involvement, or isolated extralymphatic organ involvement with distant (non-regional) nodal involvement.

Staged by _____ M.D.
_____ Registrar
Date _____

Histopathologic Type

Hodgkin's disease is divided into four major histologic types and "unclassified." These types should be recorded because they have prognostic significance. They are:

Nodular sclerosis
Lymphocyte predominance
Mixed cellularity
Lymphocyte depletion
Unclassified

Histologic classification should be based on paraffin-embedded hematoxylin and eosin-stained sections.

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Non-Hodgkin's Lymphoma

The histologic classification of the non-Hodgkin's lymphomas has been an area of considerable controversy. Currently, various competing classifications are in use, including those of Rappaport, Lukes and Collins, WHO, Dorfman, Kiel, and the British National Lymphoma Investigation Group. In an effort to bring some uniformity to the classification of these disorders, an international panel of expert pathologists has generated a Working Formulation, which attempts to provide a means of interpretation of these somewhat divergent classification schemes. This formulation provides a useful format in which to discuss the staging and workup of these lymphomas.

The anatomic staging system currently used was developed for Hodgkin's disease and has been extended to the non-Hodgkin's lymphomas, although it is more directly applicable to Hodgkin's disease. As a result, some difficulties arise in some instances when attempting to apply traditional staging systems to non- Hodgkin's lymphomas. However, in the main it has proved to be a workable system and has the advantage of being similar to that used in Hodgkin's disease and thus familiar.

The TNM classification is not a workable system for staging the malignant lymphomas, however. The site of origin of these diseases is often unclear, and there is no way to differentiate among T, N, and M. In non-Hodgkin's lymphomas, the pattern of node involvement (follicular versus diffuse) and the bulk of disease at individual sites is often more important than anatomic considerations.

ANATOMY

The major lymphatic structures include groups and chains of lymph nodes, the spleen, thymus, Waldeyer's ring, appendix, and Peyer's patches. Minor lymphoid collections are widely dispersed in other viscera and tissues, such as the bone marrow, liver, skin, bone, lung, pleura, and gonads. Involvement of extranodal sites is more commonly seen in the non-Hodgkin's lymphomas than in Hodgkin's disease.

RULES FOR CLASSIFICATION

The diagnosis of malignant lymphoma requires the biopsy of lymph nodes or of an extranodal lymphoid tumor. Frozen sections are never to be used as a definitive diagnostic source, and confirmation rests on the review of the fixed specimen.

Clinical Staging. Staging generally involves a combination of clinical, radiologic, and surgical procedures, progressing sequentially from less invasive to more invasive, necessary to define the final stage and to provide a sound basis for planning and monitoring therapy. Clinical staging includes a carefully recorded medical history, physical examination, urinalysis, chest roentgenography, blood chemistry studies, complete blood examination, and bilateral biopsies of the bone marrow. In addition, most investigators use abdominal computed tomography (CT) scan to fulfill the mandatory staging requirements. Other procedures often useful in full staging of patients include bone roentgenography, technetium 99m-labeled polyphosphate bone scans, or CT scans of the thorax (if the initial chest x-ray is abnormal). Additional procedures helpful under certain circumstances include upper GI series (if Waldeyer's ring is involved or if the patient has GI symptoms), lumbar puncture (for patients with diffuse histologies and bone marrow involvement), ultrasonography, gallium scans, and radioisotopic scans of the spleen and liver. Increasingly, surface marker studies and studies of immunoglobulin gene rearrangement have been used to characterize these lymphomas, although these presently must be thought of as research tools.

Pathologic Staging. Initial diagnosis is almost always made by surgical biopsy. In addition, biopsy of accessible extranodal primary tumors is desirable. Extranodal sites of disease at presentation are seen in about 30% of patients. About 25% of patients with non-Hodgkin's lymphomas present with evidence of abdominal disease requiring laparotomy for diagnosis. However, staging laparotomy is not routinely used in this disease and should only be used when treatment changes would be indicated from the results of the surgery. If liver involvement

is suspected, it may be biopsied by a percutaneous needle procedure, or multiple directed biopsies of both lobes may be obtained using peritoneoscopy. Although a staging laparotomy is used selectively and only after careful consideration of its impact on both staging and subsequent therapy, when used it should include splenectomy, wedge liver biopsy, and biopsies of the perisplenic, mesenteric, portahepatic, para-aortic, and bilateral iliac nodes, unless underlying medical problems prohibit such biopsies.

Retreatment Evaluation. Suspected recurrence or relapses require biopsy confirmation, particularly if a complete remission of greater than 1 year has occurred. Patients may be reevaluated for extent of disease at this juncture, using the procedures previously outlined for staging.

STAGE GROUPING

Stage I	Involvement of a single lymph node region (I) or localized involvement of a single extralymphatic organ or site (I_E).
Stage II	Involvement of two or more lymph node regions on the same side of the diaphragm (II), or localized involvement of a single associated extralymphatic organ or site and its regional nodes with or without other lymph node regions on the same side of the diaphragm (II_E).
	Note: The number of lymph node regions involved may be indicated by a subscript (e.g., II_3).
Stage III	Involvement of lymph node regions on both sides of the diaphragm (III) that may also be accompanied by localized involvement of an extralymphatic organ or site (III_E), by involvement of the spleen (III_S), or both (III_{E+S}).
Stage IV	Disseminated (multifocal) involvement of one or more extralymphatic organs with or without associated lymph node involvement, or isolated extralymphatic organ involvement with distant (nonregional) nodal involvement.

SYSTEMIC SYMPTOMS

Systemic symptoms are not as commonly associated with the non-Hodgkin's lymphomas as with Hodgkin's disease, and patients with non-Hodgkin's lymphomas often have remarkably few symptoms, even though many node areas and/or extranodal sites are involved. However, when systemic symptoms are seen, they do have prognostic significance.

Each stage is subdivided into "A" and "B" categories: "B" for those with defined systematic symptoms and "A" for those without. The B designation is given to those patients with (1) unexplained loss of more than 10% of body weight in the 6 months before diagnosis; (2) unexplained fever with temperatures above 38 C; and (3) drenching night sweats. Pruritus alone does not qualify for B classification, nor does a short febrile illness associated with an infection.* In addition, an accurate assessment of the performance status (ECOG or Karnofsky) with allowances for unrelated diseases is most important.

**Note:* Pruritus as a systemic symptom remains controversial. This symptom is hard to define quantitatively and uniformly, but when it is recurrent, generalized, and otherwise unexplained, and when it ebbs and flows parallel to disease activity, it may be the equivalent of a B symptom.

GENERAL CONSIDERATIONS

The anatomic extent of disease in the non-Hodgkin's lymphomas is defined by the appropriate sequence of diagnostic procedures selected for a given histologic subset and a particular individual. The exact sequence of staging procedures and the magnitude of invasive staging will rest on the patient's histology, the therapeutic approach contemplated, and the stage of disease. No invasive staging procedure should be used merely to change the patient's stage, if that change of stage will not alter the therapy selected or the outcome of treatment. There is always some variation—often with good reason—in the degree of completeness and adequacy of the data used for final staging.

In general, the yield from particular staging procedures depends on the histology of the patient's lymphoma. For instance, in the low-grade or indolent follicular lymphomas (see Histopathology), some 80% to 90% of patients will have positive lymphangiograms, 40% will have liver involvement, and more than 40% will have bone marrow involvement as well. When comprehensive staging is done on these patients, over 90% have Stage III–IV disease. This high frequency of advanced disease makes staging laparotomy rarely, if ever, required in the workup of follicular lymphoma, because treatment decisions are rarely influenced by the findings in the majority of patients.

In contrast, in the intermediate- or high-grade lymphomas, a much lower incidence of visceral disease is generally found at initial staging. As an example, some 30% to 40% of patients have positive lymphangio-

grams, the frequency of positive bone marrows is about 15% to 20%, and about 15% to 20% of liver biopsies are positive. After final comprehensive staging, about 25% to 30% of patients with diffuse aggressive lymphoma appear to have localized (Stage I and II) disease. Again, the importance of the extent of staging rests on the subsequent therapeutic approaches taken and the success of that therapy. Comprehensive staging is required if a localized form of therapy (i.e., involved field irradiation) is being considered.

CT scans are a useful addition to the staging procedures. They should be done before lymphangiography, because after lymphangiography the increase in size of nodes may lead to a false CT. Moreover, foci of lymphoreticular disease in the para-aortic region above the level of the second lumbar vertebra, in the portahepatic, splenic hilus, mesentery, gut wall, and retrocrural nodes, and in other sites in the abdomen cannot be demonstrated by lymphangiography. On the other hand, CT scanning is unable to detect small defects in otherwise normal-sized nodes. Thus, a complementary role of CT scanning and lymphangiography is seen in the non-Hodgkin's lymphomas.

HISTOPATHOLOGIC TYPE

Although individual institutions and particular pathologists may use one of the many classifications of these lymphomas mentioned earlier (see Introduction), the corresponding Working Formulation equivalent should be identified so that interinstitutional comparisons can be made and accurate staging approaches selected. The Working Formulation is listed below. It should be noted that the term non-Hodgkin's lymphoma is not used, follicular is used rather than nodular, and surface markers are not required.

HISTOPATHOLOGIC GRADE (G)

Working Formulation

I. Low-grade malignant lymphoma
   - A. Small lymphocytic
   - B. Follicular, predominantly small cleaved cell
   - C. Follicular mixed, small and large cell

II. Intermediate-grade malignant lymphoma
   - D. Follicular, predominantly large cell
   - E. Diffuse small cleaved cell
   - F. Diffuse mixed, small and large cell
   - G. Diffuse large cell, cleaved or noncleaved

III. High-grade malignant lymphoma
   - H. Diffuse large cell immunoblastic
   - I. Lymphoblastic (convoluted or nonconvoluted)
   - J. Small noncleaved cell (Burkitt's or non-Burkitt's)

IV. Miscellaneous
   - Composite
   - Mycosis fungoides
   - Other

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Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	DEFINITIONS
		Stage Grouping
[ ]	[ ]	Stage I Involvement of single lymph node region (I) or localized involvement of a single extralymphatic organ or site (I_E).
[ ]	[ ]	Stage II Involvement of two or more lymph node regions on the same side of the diaphragm (II), or localized involvement of a single associated extralymphatic organ or site and its regional nodes with or without other lymph node regions on the same side of the diaphragm (II_E). NOTE: The number of lymph node regions involved may be indicated by a subscript (e.g., II_3).
[ ]	[ ]	Stage III Involvement of lymph node regions on both sides of the diaphragm (III), which may also be accompanied by localized involvement of an extralymphatic organ or site (III_E), by involvement of the spleen (III_S), or both (III_{E+S}).
[ ]	[ ]	Stage IV Disseminated (multifocal) involvement of one or more extralymphatic organs, with or without associated lymph node involvement, or isolated extralymphatic organ involvement with distant (nonregional) nodal involvement.

Staged by _____ M.D.
_____ Registrar
Date _____

Histopathologic Type

While individual institutions and particular pathologists may use one of the many classifications of these lymphomas mentioned earlier (see Introduction), the corresponding Working Formulation equivalent should be identified so that interinstitutional comparisons can be made and accurate staging approaches selected. The modified Working Formulation is listed below. It should be noted that the term *non-Hodgkin's lymphoma* is not used, *follicular* is employed rather than *nodular*, and surface markers are not required.

Histopathologic Grade (G)

Working Formulation

I. Low-Grade Malignant Lymphoma
  A. Small lymphocytic
  B. Follicular, predominantly small cleaved cell
  C. Follicular mixed, small and large cell
II. Intermediate-Grade Malignant Lymphoma
  D. Follicular, predominantly large cell
  E. Diffuse small cleaved cell
  F. Diffuse mixed, small and large cell
  G. Diffuse large cell, cleaved/noncleaved
III. High-Grade Malignant Lymphoma
  H. Diffuse large cell immunoblastic
  I. Small noncleaved cell (Burkitt's/non-Burkitt's)
  J. Lymphoblastic (convoluted/nonconvoluted)
IV. Miscellaneous
  Composite
  Mycosis fungoides
  Other

PEDIATRIC CANCERS

Pediatric tumors are classified according to the recommendations of the Societe Internationale d'Oncologie Pediatrique (SIOP). The TNM classification used in this edition of the Manual for pediatric tumors is the same as that published in the 1988 edition. Tumors are staged clinically before definitive treatment and pathologically after examination of the resected specimen. The prognosis of childhood cancers has improved dramatically in the last 15 years. In clinical trials and cooperative group protocols, a different or modified staging classification may be used.

Malignant tumors of childhood include neuroblastomas, nephroblastomas or Wilms' tumor, and the soft tissue sarcomas, which include the rhabdomyosarcomas. Neuroblastomas are the most common tumor found at birth. Nephroblastomas have had over 75 synonyms, which will not be listed. Rhabdomyosarcoma is the most common soft tissue sarcoma of childhood.

These pediatric cancers may be present at birth or may develop during the first several years of life. Some pediatric cancers, especially Wilms' tumor, may be associated with congenital anomalies in other organs. Cancers in children are staged the same as in adults, except in one respect. For children, it is necessary to include a category for those cases in which a surgical exploration was carried out and a nonresectable tumor found. Such cases are designated with a "c" in the T category; for example, pT3c means that a nonresectable tumor was found on surgical exploration. The other two staging elements—that is, the N and M—are completed and the stage assigned according to all three categories.

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Nephroblastoma (Wilms' Tumor)

C64.9 Kidney

Nephroblastoma, or Wilms' tumor, most commonly occurs in the kidney of young children, sometimes bilaterally. Histologically, these tumors are often mixed; that is, composed of stromal and epithelial derivatives in various stages of differentiation. Nephroblastomas are most commonly seen in children under age 8 years, with peak incidence occurring in the second year of life. Bilateral and familial nephroblastomas tend to occur at a younger age than nephroblastomas in general. The younger the child, the better the prognosis. Nephroblastomas typically present as an abdominal mass; plasma and urine erythropoietin levels are commonly elevated. Treatment for these cancers has improved dramatically in the past 15 years. These tumors are staged clinically and pathologically.

ANATOMY

Primary Site. Nephroblastomas arise from the kidneys. These tumors may be bilateral and multiple.

Regional Lymph Nodes. The regional lymph nodes are: the hilar nodes, the para-aortic nodes, and the paracaval nodes located between the diaphragm and the bifurcation of the aorta. All other lymph nodes involved are considered distant metastases and must be coded as M1.

Metastatic Sites. Distant metastases are most common in the lungs, liver, and regional lymph nodes. Tumor may also extend along the renal vein and the inferior vena cava. Involvement of the opposite kidney is classified as T4.

RULES FOR CLASSIFICATION

This classification applies only to nephroblastoma (Wilms' tumor).

Clinical Staging. Clinical classification is based on the surface area of the primary tumor as revealed by imaging, whether tumor occurs bilaterally or unilaterally, and whether or not the tumor has broken through and ruptured its capsule. Extension of the tumor through its cap-

sule worsens the prognosis. Clinical classification is based on evidence acquired from clinical, radiologic, endoscopic, and other relevant studies prior to the decision about definitive treatment. When TNM is used without a prefix, it implies clinical classification (cTNM).

Pathologic Staging. Pathologic classification is based on regional extension beyond the confines of the kidney. This is determined by evidence acquired prior to the decision about definitive treatment and is supplemented or modified by the additional evidence acquired from definitive surgery and from the examination of the resected specimen.

DEFINITION OF TNM

Clinical Classification (cTNM)

Primary Tumor (cT)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
T1 Unilateral tumor 80 cm^2 or less in area (including the kidney)*
T2 Unilateral tumor more than 80 cm^2 in area (including the kidney)*
T3 Unilateral tumor rupture before treatment
T4 Bilateral tumors

**Note:* The area is calculated by multiplying the vertical and horizontal dimensions of the radiologic shadow of the tumor and kidney.

Regional Lymph Nodes (cN)

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

Distant Metastasis (cM)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

Pathologic Classification (pTNM)

Primary Tumor (pT)

pTX Primary tumor cannot be assessed
pT0 No evidence of primary tumor
pT1 Intrarenal tumor completely encapsulated; excision complete and margins histologically free
pT2 Tumor invades beyond the capsule or renal parenchyma*; excision complete
pT3 Tumor invades beyond the capsule or renal parenchyma*; excision incomplete or preoperative or operative rupture
pT3a Microscopic residual tumor limited to the tumor bed
pT3b Macroscopic residual tumor or spillage or malignant ascites
pT3c Surgical exploration; tumor not resected
pT4 Bilateral tumors

**Note:* This includes breach of the renal capsule or tumor seen microscopically outside the capsule; tumor adhesions microscopically confirmed; infiltrations of or tumor thrombus within the renal vessels outside the kidney; and infiltration of the renal pelvis and/or ureter, peripelvic, and pericalyceal fat.

Regional Lymph Nodes (pN)

pNX Regional lymph nodes cannot be assessed
pN0 No regional lymph node metastasis
pN1 Regional lymph node metastasis
pN1a Regional lymph node metastasis completely resected
pN1b Regional lymph node metastasis incompletely resected

Distant Metastasis (pM)

pMX Presence of distant metastasis cannot be assessed
pM0 No distant metastasis
pM1 Distant metastasis

CLINICAL STAGE GROUPING (cTNM)

Stage I	T1	N0	M0
Stage II	T2	N0	M0
Stage III	T1	N1	M0
	T2	N1	M0
	T3	Any N	M0
Stage IVA	T1	Any N	M1
	T2	Any N	M1
	T3	Any N	M1
Stage IVB	T4	Any N	Any M

PATHOLOGIC STAGE GROUPING (pTNM)

Stage I	pT1	pN0	M0*
Stage II	pT1	pN1a	M0
	pT2	pN0	M0
	pT2	pN1a	M0
Stage IIIA	pT3a	pN0	M0
	pT3a	pN1a	M0
Stage IIIB	pT1	pN1b	M0
	pT2	pN1b	M0
	pT3a	pN1b	M0
	pT3b	Any pN	M0
	pT3c	Any pN	M0

(continued on next page)

Stage IVA	pT1	Any pN	M1
	pT2	Any pN	M1
	pT3a	Any pN	M1
	pT3b	Any pN	M1
	pT3c	Any pN	M1
Stage IVB	pT4	Any pN	Any M

**Note:* For pathologic stage grouping, a clinical M is acceptable.

HISTOPATHOLOGIC TYPE

These are a distinctive group of tumors that show various histologies, differentiation, and components. A number of synonyms include angiomyosarcoma, adenosarcoma, mesoblastic nephroma, and embryoma. The various synonyms, of which there are over 75, reflect the different tissue components that may be present.

NATIONAL WILMS' TUMOR STUDY GROUP (NWTSG)

Most children with Wilms' tumor treated in the United States are staged and treated on the basis of the NWTSG protocol. The pathologic stage grouping is identical to that of the AJCC, with the only exception being a separate category (Stage IV) for bilateral Wilms' tumor.

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Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age ____ Sex ____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	DEFINITIONS
		Primary Tumor (T)
[ ]	[ ]	TX Primary tumor cannot be assessed
[ ]	[ ]	T0 No evidence of primary tumor
[ ]	[ ]	T1 Unilateral tumor 80 cm^2 or less in area (including kidney)*
[ ]	[ ]	T2 Unilateral tumor more than 80 cm^2 in area (including kidney)*
[ ]	[ ]	T3 Unilateral tumor rupture before treatment
[ ]	[ ]	T4 Bilateral tumors
		* *Note:* The area is calculated by multiplying the vertical and horizontal dimensions of the radiologic shadow of the tumor and kidney.
		Lymph Node (N)
[ ]	[ ]	NX Regional lymph nodes cannot be assessed
[ ]	[ ]	N0 No regional lymph node metastasis
[ ]	[ ]	N1 Regional lymph node metastasis
		Distant Metastasis (M)
[ ]	[ ]	MX Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0 No distant metastasis
[ ]	[ ]	M1 Distant metastasis
		Primary Tumor (pT)
[ ]	[ ]	pTX Primary tumor cannot be assessed
[ ]	[ ]	pT0 No evidence of primary tumor
[ ]	[ ]	pT1 Intrarenal tumor completely encapsulated; excision complete and margins histologically free
[ ]	[ ]	pT2 Tumor invades beyond the capsule or renal parenchyma;* excision complete
[ ]	[ ]	pT3 Tumor invades beyond the capsule or renal parenchyma;* excision incomplete *or* preoperative or operative rupture
[ ]	[ ]	pT3a Microscopic residual tumor limited to tumor bed
[ ]	[ ]	pT3b Macroscopic residual tumor or spillage or malignant ascites
[ ]	[ ]	pT3c Surgical exploration only, tumor not resected
[ ]	[ ]	pT4 Bilateral tumors
		* *Note:* This includes breach of the renal capsule or tumor seen microscopically outside the capsule; tumor adhesions microscopically confirmed; infiltrations of or tumor thrombus within the renal vessels outside the kidney; and infiltration of the renal pelvis and/or ureter, peripelvic, and pericalyceal fat.
		Lymph Node (pN)
[ ]	[ ]	pNX Regional lymph nodes cannot be assessed
[ ]	[ ]	pN0 No regional lymph node metastasis
[ ]	[ ]	pN1 Regional lymph node metastasis
[ ]	[ ]	pN1a Regional lymph node metastasis completely resected
[ ]	[ ]	pN1b Regional lymph node metastasis incompletely resected
		Distant Metastasis (pM)
[ ]	[ ]	pMX Presence of distant metastasis cannot be assessed
[ ]	[ ]	pM0 No distant metastasis
[ ]	[ ]	pM1 Distant metastasis

(continued on next page)

Clin	Path				
		Clinical Stage Grouping (cTNM)			
[ ]	[ ]	I	T1	N0	M0
[ ]	[ ]	II	T2	N0	M0
[ ]	[ ]	III	T1	N1	M0
			T2	N1	M0
			T3	Any N	M0
[ ]	[ ]	IVA	T1	Any N	M1
			T2	Any N	M1
			T3	Any N	M1
[ ]	[ ]	IVB	T4	Any N	Any M
		Pathologic Stage Grouping (pTNM)			
[ ]	[ ]	I	pT1	pN0	M0
[ ]	[ ]	II	pT1	pN1a	M0
			pT2	pN0	M0
			pT2	pN1a	M0
[ ]	[ ]	IIIA	pT3a	pN0	M0
			pT3a	pN1a	M0
[ ]	[ ]	IIIB	pT1	pN1b	M0
			pT2	pN1b	M0
			pT3a	pN1b	M0
			pT3b	Any pN	M0
			pT3c	Any pN	M0
[ ]	[ ]	IVA	pT1	Any pN	M1
			pT2	Any pN	M1
			pT3a	Any pN	M1
			pT3b	Any pN	M1
			pT3c	Any pN	M1
[ ]	[ ]	IVB	pT4	Any pN	Any M

Staged by _____ M.D.

_____ Registrar

Date _____

Histopathologic Type

These are a distinctive group of tumors that show various histologies, differentiation, and components. A number of synonyms include angiomyosarcoma, adenosarcoma, mesoblastic nephroma, and embryoma. The various synonyms, of which there are over 75, reflect the different tissue components that may be present.

50

Neuroblastoma

Neuroblastomas usually arise from the adrenal glands. These tumors are highly malignant, with a 5-year survival of approximately 30% when discovered in the first year of life. Spontaneous regression of neuroblastomas does occur, especially in very young infants. For this reason, these tumors are of great interest to oncologists and medical scientists. Neuroblastomas are almost always found in children under age 8 years. These tumors may elaborate epinephrine and norepinephrine. Neuroblastomas can cause widespread and rapid metastases. Clinical manifestations vary, depending on the tumor site and extent. In addition to staging, a number of prognostic factors have been identified for neuroblastoma. As a prognostic indicator, amplification of the *c-myc* oncogene is under extensive study. In the future, the extent of amplification may be incorporated formally into the staging system.

ANATOMY

Primary Site. Neuroblastomas usually originate in the adrenal medulla. However, they may be found at other sites; for example, in the posterior mediastinum or anywhere along the course of the sympathetic chain, from the cervical region to the pelvis. These tumors may be multicentric in origin.

Regional Lymph Nodes. The regional lymph nodes are defined as follows:

Cervical region: cervical and supraclavicular nodes
Thoracic region: intrathoracic and infraclavicular nodes
Abdominal and pelvic regions: subdiaphragmatic, intra-abdominal, and pelvic nodes, including the external iliac nodes
Other regions: the appropriate regional lymph nodes

Metastatic Sites. Metastases are usually found in the liver, orbit, and bones, although nearly every organ can be affected. When the tumor develops in utero, the placenta may also be involved.

RULES FOR CLASSIFICATION

Clinical Staging. Because it is often impossible to differentiate between the primary tumor and the adjacent lymph nodes, the T assessment relates to the total mass. When there is doubt about multicentricity and metastasis, the latter is presumed. Size is estimated clinically or radiologically; for classification, the larger measurement should be used. There should be histologic confirmation of the disease and/or confirmation by biochemical tests.

Pathologic Staging. All clinical data and that found on examination of the surgically resected specimen is to be used. Definitions of pTNM differ from cTNM.

DEFINITION OF TNM

Clinical Classification (cTNM)

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
T1 Single tumor 5 cm or less in greatest dimension
T2 Single tumor more than 5 cm but not more than 10 cm in greatest dimension
T3 Single tumor more than 10 cm in greatest dimension
T4 Multicentric tumors occurring simultaneously

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

DEFINITION OF TNM

Pathologic Classification (pTNM)

Primary Tumor (pT)

pTX Primary tumor cannot be assessed
pT0 No evidence of primary tumor
pT1 Excision of tumor complete and margins histologically free
pT2 The category does not apply to neuroblastoma
pT3 Residual tumor
- pT3a Microscopic residual tumor
- pT3b Macroscopic residual tumor or grossly incomplete excision
- pT3c Surgical exploration tumor not resected

pT4 Multicentric tumors

Regional Lymph Nodes (pN)

pNX Regional lymph nodes cannot be assessed
pN0 No regional lymph node metastasis
pN1 Regional lymph node metastasis
- pN1a Regional lymph node metastases completely resected
- pN1b Regional lymph node metastases incompletely resected

Distant Metastasis (pM)

pMX Presence of distant metastasis cannot be assessed
pM0 No distant metastasis
pM1 Distant metastasis

CLINICAL STAGE GROUPING (cTNM)

Stage I	T1	N0	M0
Stage II	T2	N0	M0
Stage III	T1	N1	M0
	T2	N1	M0
	T3	Any N	M0
Stage IVA	T1	Any N	M1
	T2	Any N	M1
	T3	Any N	M1
Stage IVB	T4	Any N	Any M

PATHOLOGIC STAGE GROUPING (pTNM)

Stage I	pT1	pN0	M0*
Stage II	pT1	pN1a	M0
Stage IIIA	pT3a	pN0	M0
	pT3a	pN1a	M0
Stage IIIB	pT1	pN1b	M0
	pT3a	pN1b	M0
	pT3b	Any pN	M0
	pT3c	Any pN	M0
Stage IVA	pT1	Any pN	M1
	pT3a	Any pN	M1
	pT3b	Any pN	M1
	pT3c	Any pN	M1
Stage IVB	pT4	Any pN	Any M

*Note: For pathologic stage grouping, a clinical M is acceptable.

HISTOPATHOLOGIC TYPE

Depending on the extent of cellular differentiation, these tumors can be designated by several terms, including sympathicoblastomas, sympathicogoniomas, malignant ganglioneuromas, and gangliosympathicoblastomas. Ganglioneuroma, apparently a well-differentiated neuroblastoma, is also covered by this staging classification, even though it behaves in a benign manner.

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Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

Clin	Path	DEFINITIONS
		Primary Tumor (T)
[ ]	[ ]	TX Primary tumor cannot be assessed
[ ]	[ ]	T0 No evidence of primary tumor
[ ]	[ ]	T1 Single tumor 5 cm or less in greatest dimension
[ ]	[ ]	T2 Single tumor more than 5 cm but not more than 10 cm in greatest dimension
[ ]	[ ]	T3 Single tumor more than 10 cm in greatest dimension
[ ]	[ ]	T4 Multicentric tumors occurring simultaneously
		Lymph Node (N)
[ ]	[ ]	NX Regional lymph nodes cannot be assessed
[ ]	[ ]	N0 No regional lymph node metastasis
[ ]	[ ]	N1 Regional lymph node metastasis
		Distant Metastasis (M)
[ ]	[ ]	MX Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0 No distant metastasis
[ ]	[ ]	M1 Distant metastasis
		Primary Tumor (pT)
[ ]	[ ]	pTX Primary tumor cannot be assessed
[ ]	[ ]	pT0 No evidence of primary tumor
[ ]	[ ]	pT1 Excision of tumor complete and margins histologically free
[ ]	[ ]	pT2 The category does not apply to neuroblastoma
[ ]	[ ]	pT3 Residual tumor
[ ]	[ ]	pT3a Microscopic residual tumor
[ ]	[ ]	pT3b Macroscopic residual tumor or grossly incomplete excision
[ ]	[ ]	pT3c Surgical exploration tumor not resected
[ ]	[ ]	pT4 Multicentric tumors
		Lymph Node (pN)
	[ ]	pNX Regional lymph nodes cannot be assessed
	[ ]	pN0 No regional lymph node metastasis
	[ ]	pN1 Regional lymph node metastasis
	[ ]	pN1a Regional lymph node metastases completely resected
	[ ]	pN1b Regional lymph node metastases incompletely resected
		Distant Metastasis (pM)
[ ]	[ ]	pMX Presence of distant metastasis cannot be assessed
[ ]	[ ]	pM0 No distant metastasis
[ ]	[ ]	pM1 Distant metastasis

(continued on next page)

Clin	Path				
		Clinical Stage Grouping (cTNM)			
[ ]	[ ]	I	T1	N0	M0
[ ]	[ ]	II	T2	N0	M0
[ ]	[ ]	III	T1	N1	M0
			T2	N1	M0
			T3	Any N	M0
[ ]	[ ]	IVA	T1	Any N	M1
			T2	Any N	M1
			T3	Any N	M1
[ ]	[ ]	IVB	T4	Any N	Any M
		Pathologic Stage Grouping (pTNM)			
[ ]	[ ]	I	pT1	pN0	M0
[ ]	[ ]	II	pT1	pN1a	M0
[ ]	[ ]	IIIA	pT3a	pN0	M0
			pT3a	pN1a	M0
[ ]	[ ]	IIIB	pT1	pN1b	M0
			pT3a	pN1b	M0
			pT3b	Any pN	M0
			pT3c	Any pN	M0
[ ]	[ ]	IVA	pT1	Any pN	M1
			pT2	Any pN	M1
			pT3a	Any pN	M1
			pT3b	Any pN	M1
			pT3c	Any pN	M1
[ ]	[ ]	IVB	pT4	Any pN	Any M

Staged by _____ M.D.

_____ Registrar

Date _____

Histopathologic Type

These tumors can be designated by several terms, including sympathicoblastomas, sympathicogoniomas, malignant ganglioneuromas, and gangliosympathicoblastomas, depending on the extent of cellular differentiation. Ganglioneuroma, which apparently is a well differentiated neuroblastoma, is also covered by this staging classification, even though it behaves in a benign fashion.

51

Soft-Tissue Sarcoma—Pediatric

Malignant soft-tissue tumors can occur in infants and in children. Found in many sites, these tumors include various histologic types. The most important is the embryonic rhabdomyosarcoma, or sarcoma botryoides, which can arise in numerous organs. Some histologic types of sarcomas are found only in children. These tumors commonly have an embryonic appearance histologically, and usually are highly malignant. These tumors can be staged clinically and pathologically.

ANATOMY

Primary Site. Soft-tissue sarcomas can involve nearly all anatomic sites. In children, these tumors may even affect unusual sites, such as the vagina or extrahepatic bile ducts, which are rarely involved in adults.

The primary tumor site should be indicated according to the following notations:

ORB Orbit
HEA Head and neck
LIM Limbs
PEL **Pelvis (including** walls, genital tract, and viscera)
ABD Abdomen (including walls and viscera)
THO Thorax (including walls, diaphragm, and viscera)
OTH Other

Regional Lymph Nodes. The regional lymph nodes are those appropriate to the location of the primary tumor, as in the following:

Head and neck: cervical and supraclavicular lymph nodes
Abdominal and pelvic: subdiaphragmatic, intra-abdominal, and ilioinguinal lymph nodes
Upper limbs: ipsilateral epitrochlear and axillary lymph nodes
Lower limbs: ipsilateral popliteal and inguinal lymph nodes

In the case of unilateral tumors, all contralateral involved lymph nodes are considered distant metastases and should be coded as M1.

Metastatic Sites. Because these tumors are found in many sites, they can involve numerous organs either by direct extension or by distant spread, usually through the bloodstream.

RULES FOR CLASSIFICATION

Clinical Staging. There is a clinical and pathologic TNM classification for pediatric soft-tissue tumors. Clinical staging is based on clinical examination, including imaging and laboratory studies.

Pathologic Staging. Pathologic classification is based on information obtained from pretreatment clinical classification and from surgery and pathologic examination of the resected specimen.

The classification for soft-tissue sarcomas is designed to apply primarily to rhabdomyosarcomas in childhood, but it may also be used for other soft-tissue sarcomas. In rhabdomyosarcoma, bone marrow examination is recommended.

There should be histologic verification of the disease.

DEFINITION OF TNM

Clinical Classification (cTNM)

Primary Tumor (T)

TX Primary tumor cannot be assessed
T0 No evidence of primary tumor
T1 Tumor limited to the organ or tissue of origin
  T1a Tumor 5 cm or less in greatest dimension
  T1b Tumor more than 5 cm in greatest dimension
T2 Tumor invades contiguous organ(s) or tissue(s) and/or with adjacent malignant effusion
  T2a Tumor 5 cm or less in greatest dimension
  T2b Tumor more than 5 cm in greatest dimension

Note: The categories T3 and T4 do not apply. The existence of more than one tumor generally is considered as a primary tumor with distant metastases.

Regional Lymph Nodes (N)

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

Distant Metastasis (M)

MX Presence of distant metastasis cannot be assessed
M0 No distant metastasis
M1 Distant metastasis

DEFINITION OF TNM

Pathologic Classification (pTNM)

Primary Tumor (pT)

pTX Primary tumor cannot be assessed
pT0 No evidence of primary tumor
pT1 Tumor limited to the organ or tissue of origin; excision complete and margins histologically free
pT2 Tumor invades beyond the organ or tissue of origin; excision complete and margins histologically free
pT3 Tumor invades beyond the organ or tissue of origin; excision incomplete
- pT3a Microscopic residual tumor
- pT3b Macroscopic residual tumor or adjacent malignant effusion
- pT3c Surgical exploration tumor not resected

Regional Lymph Nodes (pN)

pNX Regional lymph nodes cannot be assessed
pN0 No regional lymph node metastasis
pN1 Regional lymph node metastasis
- pN1a Regional lymph node metastasis completely resected
- pN1b Regional lymph node metastasis incompletely resected

Distant Metastasis (pM)

pMX Presence of distant metastasis cannot be assessed
pM0 No distant metastasis
pM1 Distant metastasis

CLINICAL STAGE GROUPING (cTNM)

It is recommended that the full TNM classification always be used.

Stage I	T1a	N0	M0
	T1b	N0	M0
Stage II	T2a	N0	M0
	T2b	N0	M0
Stage III	Any T	N1	M0
Stage IV	Any T	Any N	M1

When the regional lymph nodes cannot be assessed clinically or radiologically, NX should be considered N0 in Stages I and II. Further studies are required to determine the exact significance of N0, N1, and NX in such cases as pelvic tumors.

PATHOLOGIC STAGE GROUPING (pTNM)

Stage I	pT1	pN0	M0*
Stage II	pT1	pN1a	M0
	pT2	pN0	M0
	pT2	pN1a	M0
Stage IIIA	pT3a	pN0	M0
	pT3a	pN1a	M0
Stage IIIB	pT3b	Any pN	M0
	pT3c	Any pN	M0
	Any pT	pN1b	M0
Stage IV	Any pT	Any pN	M1

**Note:* For pathologic stage grouping, a clinical M is acceptable.

HISTOPATHOLOGIC TYPE

Histology can include the soft-tissue tumors found in adults. In general, soft-tissue sarcomas are relatively rare in children. Some sarcomas—for instance, osteogenic sarcomas found in children and young adolescents—are classified under the musculoskeletal system.

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Data Form for Cancer Staging

Patient identification
Name _____
Address _____
Hospital or clinic number _____
Age _____ Sex _____ Race _____

Institution identification
Hospital or clinic _____
Address _____

Oncology Record

Anatomic site of cancer _____
Histologic type _____
Grade (G) _____
Date of classification _____

Chronology of classification
[ ] Clinical (use all data prior to first treatment)
[ ] Pathologic (if definitively resected specimen available)

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		Primary Tumor (T)
[ ]	[ ]	TX Primary tumor cannot be assessed
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[ ]	[ ]	T1 Tumor limited to the organ or tissue of origin
[ ]	[ ]	T1a Tumor 5 cm or less in greatest dimension
[ ]	[ ]	T1b Tumor more than 5 cm in greatest dimension
[ ]	[ ]	T2 Tumor invades contiguous organs or tissue(s) and/or with adjacent malignant effusion
[ ]	[ ]	T2a Tumor 5 cm or less in greatest dimension
[ ]	[ ]	T2b Tumor more than 5 cm in greatest dimension
		Lymph Node (N)
[ ]	[ ]	NX Regional lymph nodes cannot be assessed
[ ]	[ ]	N0 No regional lymph node metastasis
[ ]	[ ]	N1 Regional lymph node metastasis
		Distant Metastasis (M)
[ ]	[ ]	MX Presence of distant metastasis cannot be assessed
[ ]	[ ]	M0 No distant metastasis
[ ]	[ ]	M1 Distant metastasis
		Primary Tumor (pT)
[ ]	[ ]	pTX Primary tumor cannot be assessed
[ ]	[ ]	pT0 No evidence of primary tumor
[ ]	[ ]	pT1 Tumor limited to organ or tissue of origin; excision complete and margins histologically free
[ ]	[ ]	pT2 Tumor invades beyond the organ or tissue of origin; excision complete and margins histologically free
[ ]	[ ]	pT3 Tumor invades beyond the organ or tissue of origin; excision incomplete
[ ]	[ ]	pT3a Microscopic residual tumor
[ ]	[ ]	pT3b **Macroscopic residual tumor** or adjacent malignant effusion
[ ]	[ ]	pT3c Surgical exploration tumor not resected
		Lymph Node (pN)
[ ]	[ ]	pNX Regional lymph nodes cannot be assessed
[ ]	[ ]	pN0 No regional lymph node metastasis
[ ]	[ ]	pN1 Regional lymph node metastasis
[ ]	[ ]	pN1a Regional lymph node metastasis completely resected
[ ]	[ ]	pN1b Regional lymph node metastasis incompletely resected
		Distant Metastasis (pM)
[ ]	[ ]	pMX Presence of distant metastasis cannot be assessed
[ ]	[ ]	pM0 No distant metastasis
[ ]	[ ]	pM1 Distant metastasis

(continued on next page)

Clin	Path				
		Clinical Stage Grouping (cTNM)			
[ ]	[ ]	I	T1a	N0	M0
			T1b	N0	M0
[ ]	[ ]	II	T2a	N0	M0
			T2b	N0	M0
[ ]	[ ]	III	Any T	N1	M0
[ ]	[ ]	IV	Any T	Any N	M1
		Pathologic Stage Grouping (pTNM)			
[ ]	[ ]	I	pT1	pN0	M0
[ ]	[ ]	II	pT1	pN1a	M0
			pT2	pN0	M0
			pT2	pN1a	M0
[ ]	[ ]	IIIA	pT3a	pN0	M0
			pT3a	pN1a	M0
[ ]	[ ]	IIIB	pT3b	Any pN	M0
			pT3c	Any pN	M0
			Any pT	pN1b	M0
[ ]	[ ]	IV	Any pT	Any pN	M1

Staged by _____ M.D.

_____ Registrar

Date _____

Histopathologic Type

Histology can include the soft-tissue tumors that are found in adults. In general, soft-tissue sarcomas are relatively rare in children. Some sarcomas, for instance, osteogenic sarcomas that are found in children or young adolescents, are classified under the Musculoskeletal System.

PART III

Personnel of the American Joint Committee on Cancer

PERSONNEL OF THE AMERICAN JOINT COMMITTEE ON CANCER FULL COMMITTEE MEMBERSHIP LISTING FOR THE FOURTH EDITION (1959–1991)

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*Robert W. Beart, Jr., M.D.
*Harold E. Bowman, M.D.
David G. Bragg, M.D.
Weldon K. Bullock, M.D.
Paul R. Cannon, M.D.
Paul Carbone, M.D.
*David T. Carr, M.D. (Ex-officio)
William M. Christopherson, M.D.
Murray M. Copeland, M.D.
Michael P. Corder, M.D.
Anthony R. Curreri, M.D.
Sidney J. Cutler, Sc.D.
Herbert Derman, M.D.
Theodore P. Eberhard, M.D.
Jack Edeiken, M.D.
*Harmon J. Eyre, M.D.
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