RETINOBLASTOMA STAGING FORM

CLINICAL *Extent of disease before any treatment*	STAGE CATEGORY DEFINITIONS		PATHOLOGIC *Extent of disease through completion of definitive surgery*
❒ y clinical – staging completed after neoadjuvant therapy but before subsequent surgery	TUMOR SIZE: _____	LATERALITY: ☐ left ☐ right ☐ bilateral	❒ y pathologic – staging completed after neoadjuvant therapy AND subsequent surgery
	PRIMARY TUMOR (T)		
❑ TX	Primary tumor cannot be assessed		❑ pTX
❑ T0	No evidence of primary tumor		❑ pT0
❑ T1	Tumors no more than 2/3 the volume of the eye with no vitreous or subretinal seeding.		
pT1	Tumor confined to eye with no optic nerve or choroidal invasion.		❑ pT1
❑ T1a	No tumor in either eye is greater than 3 mm in largest dimension or located closer than 1.5 mm to the optic nerve or fovea.		
❑ T1b	At least one tumor is greater than 3 mm in largest dimension or located closer than 1.5 mm to the optic nerve or fovea. No retinal detachment or subretinal fluid beyond 5 mm from the base of the tumor.		
❑ T1c	At least one tumor is greater than 3 mm in largest dimension or located closer than 1.5 mm to the optic nerve or fovea. With retinal detachment or subretinal fluid beyond 5 mm from the base of the tumor.		
❑ T2	Tumors no more than 2/3 the volume of the eye with vitreous or subretinal seeding. Can have retinal detachment.		
pT2	Tumor with minimal optic nerve and/or choroidal invasion		❑ pT2
❑ T2a	Focal vitreous and/or subretinal seeding of fine aggregates of tumor cells is present, but no large clumps or "snowballs" of tumor cells.		
pT2a	Tumor superficially invades optic nerve head but does not extend past lamina cribrosa *or* tumor exhibits focal choroidal invasion.		❑ pT2a
❑ T2b	Massive vitreous and/or subretinal seeding is present, defined as diffuse clumps or "snowballs" of tumor cells.		
pT2b	Tumor superficially invades optic nerve head but does not extend past lamina cribrosa *and* exhibits focal choroidal invasion.		❑ pT2b
❑ T3	Severe intraocular disease		
pT3	Tumor with significant optic nerve and/or choroidal invasion		❑ pT3
❑ T3a	Tumor fills more than 2/3 of the eye.		
pT3a	Tumor invades optic nerve past lamina cribrosa but not to surgical resection line *or* tumor exhibits massive choroidal invasion.		❑ pT3a
❑ T3b	One or more complications present, which may include tumor-associated neovascular or angle closure glaucoma, tumor extension into the anterior segment, hyphema, vitreous hemorrhage, or orbital cellulitis.		❑ T3b
pT3b	Tumor invades optic nerve past lamina cribrosa but not to surgical resection line *and* exhibits massive choroidal invasion.		❑ pT3b
❑ T4	Extraocular disease detected by imaging studies.		
pT4	Tumor invades optic nerve to resection line or exhibits extraocular extension elsewhere.		❑ pT4
❑ T4a	Invasion of optic nerve.		
pT4a	Tumor invades optic nerve to resection line but no extraocular extension identified		❑ pT4a
❑ T4b	Invasion into the orbit.		
pT4b	Tumor invades optic nerve to resection line and extraocular extension identified		❑ pT4b
❑ T4c	Intracranial extension not past chiasm.		
❑ T4d	Intracranial extension past chiasm.		

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	REGIONAL LYMPH NODES (N)	
❑ NX ❑ N0 ❑ N1 ❑ N2	**Clinical** Regional lymph nodes cannot be assessed No regional lymph node involvement Regional lymph node involvement (preauricular, cervical, submandibular) Distant lymph node involvement	❑ NX ❑ N0 ❑ N1 ❑ N2
	DISTANT METASTASIS (M)	
❑ M0 ❑ M1 ❑ M1a ❑ M1b ❑ M1c ❑ M1d ❑ M1e	**Clinical** No distant metastasis (no pathologic M0; use clinical M to complete stage group) Systemic metastasis. Single lesion to sites other than CNS Multiple lesions to sites other than CNS. Prechiasmatic CNS lesion(s). Postchiasmatic CNS lesion(s). Leptomeningeal and/or CSF involvement. **Pathologic** Metastasis to sites other than CNS. Single lesion. Multiple lesions. CNS metastasis. Discrete mass(es) without leptomeningeal and/or CSF involvement. Leptomeningeal and/or CSF involvement.	❑ M1 ❑ M1a ❑ M1b ❑ M1c ❑ M1d ❑ M1e

ANATOMIC STAGE • PROGNOSTIC GROUPS

CLINICAL	PATHOLOGIC
No stage grouping is presently recommended	No stage grouping is presently recommended

PROGNOSTIC FACTORS (SITE-SPECIFIC FACTORS)

REQUIRED FOR STAGING: None

CLINICALLY SIGNIFICANT:

Extension evaluated at enucleation _____
RB gene mutation _____
Positive family history of retinoblastoma _____
Primary globe-sparing treatment failure _____
Greatest linear extent of choroid involved by choroidal tumor invasion _____

Histologic Grade (G) *(also known as overall grade)*

Grading system	*Grade*
❑ 2 grade system	❑ Grade I or 1
❑ 3 grade system	❑ Grade II or 2
❑ 4 grade system	❑ Grade III or 3
❑ No 2, 3, or 4 grade system is available	❑ Grade IV or 4

General Notes:
For identification of special cases of TNM or pTNM classifications, the "m" suffix and "y," "r," and "a" prefixes are used. Although they do not affect the stage grouping, they indicate cases needing separate analysis.

m suffix indicates the presence of multiple primary tumors in a single site and is recorded in parentheses: pT(m)NM.

y prefix indicates those cases in which classification is performed during or following initial multimodality therapy. The cTNM or pTNM category is identified by a "y" prefix. The ycTNM or ypTNM categorizes the extent of tumor actually present at the time of that examination. The "y"

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ADDITIONAL DESCRIPTORS

Lymphatic Vessel Invasion (L) and Venous Invasion (V) have been combined into Lymph-Vascular Invasion (LVI) for collection by cancer registrars. The College of American Pathologists' (CAP) Checklist should be used as the primary source. Other sources may be used in the absence of a Checklist. Priority is given to positive results.

- ❑ Lymph-Vascular Invasion Not Present (absent)/Not Identified
- ❑ Lymph-Vascular Invasion Present/Identified
- ❑ Not Applicable
- ❑ Unknown/Indeterminate

Residual Tumor (R)

The absence or presence of residual tumor after treatment. In some cases treated with surgery and/or with neoadjuvant therapy there will be residual tumor at the primary site after treatment because of incomplete resection or local and regional disease that extends beyond the limit of ability of resection.

- ❑ RX Presence of residual tumor cannot be assessed
- ❑ R0 No residual tumor
- ❑ R1 Microscopic residual tumor
- ❑ R2 Macroscopic residual tumor

categorization is not an estimate of tumor prior to multimodality therapy.

r prefix indicates a recurrent tumor when staged after a disease-free interval, and is identified by the "r" prefix: rTNM.

a prefix designates the stage determined at autopsy: aTNM.

surgical margins is data field recorded by registrars describing the surgical margins of the resected primary site specimen as determined only by the pathology report.

neoadjuvant treatment is radiation therapy or systemic therapy (consisting of chemotherapy, hormone therapy, or immunotherapy) administered prior to a definitive surgical procedure. If the surgical procedure is not performed, the administered therapy no longer meets the definition of neoadjuvant therapy.

❑ Clinical stage was used in treatment planning (describe): _____

❑ National guidelines were used in treatment planning ❑ NCCN ❑ Other (describe): _____

_____

_____

Physician signature Date/Time

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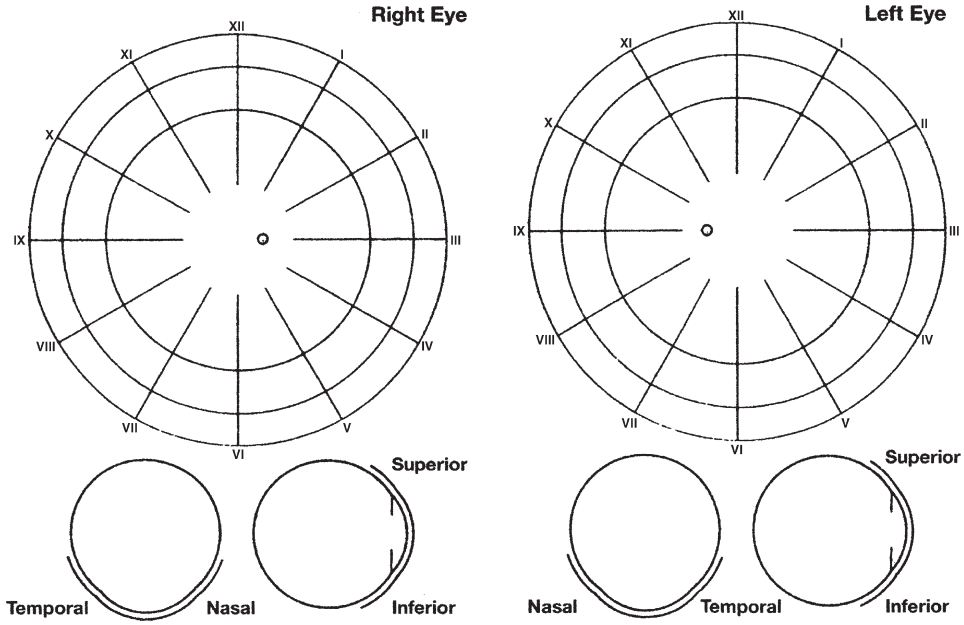
Illustration

Indicate on diagram primary tumor and regional nodes involved.



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