<table>
<thead>
<tr>
<th>Clinical</th>
<th>Stage Category Definitions</th>
<th>Pathologic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Extent of disease before any treatment</td>
<td></td>
<td>Extent of disease through completion of definitive surgery</td>
</tr>
<tr>
<td>☐ y clinical – staging completed after neoadjuvant therapy but before subsequent surgery</td>
<td>☐ y pathologic – staging completed after neoadjuvant therapy AND subsequent surgery</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Tumor Size: ________________</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Laterality: ☐ left ☐ right ☐ bilateral</td>
<td></td>
</tr>
</tbody>
</table>

### Primary Tumor (T)

- **TX**: Primary tumor cannot be assessed
- **T0**: No evidence of primary tumor
- **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.
- **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
- **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.
- **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.
- **T3**: Severe intraocular disease
- **pT3**: Tumor with significant optic nerve and/or choroidal invasion
- **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.
- **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.
- **pT3b**: Tumor invades optic nerve past lamina cribrosa but not to surgical resection line and exhibits massive choroidal invasion.
- **T4**: Extraocular disease detected by imaging studies.
- **pT4**: Tumor invades optic nerve to resection line or exhibits extraocular extension elsewhere.
- **T4a**: Invasion of optic nerve.
- **pT4a**: Tumor invades optic nerve to resection line but no extraocular extension identified
- **T4b**: Invasion into the orbit.
- **pT4b**: Tumor invades optic nerve to resection line and extraocular extension identified
- **T4c**: Intracranial extension not past chiasm.
- **pT4c**: Intracranial extension past chiasm.
# Retinoblastoma Staging Form

## Regional Lymph Nodes (N)
- **Clinical**
  - NX: Regional lymph nodes cannot be assessed
  - N0: No regional lymph node involvement
  - N1: Regional lymph node involvement (preauricular, cervical, submandibular)
  - N2: Distant lymph node involvement

## Distant Metastasis (M)
- **Clinical**
  - M0: No distant metastasis (no pathologic M0; use clinical M to complete stage group)
  - M1: Systemic metastasis
  - M1a: Single lesion to sites other than CNS
  - M1b: Multiple lesions to sites other than CNS
  - M1c: Prechiasmatic CNS lesion(s)
  - M1d: Postchiasmatic CNS lesion(s)
  - M1e: 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

## Anatomic Stage • Prognostic Groups

### Clinical
- No stage grouping is presently recommended

### Pathologic
- 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**
  - 2 grade system
  - 3 grade system
  - 4 grade system
  - No 2, 3, or 4 grade system is available

- **Grade**
  - Grade I or 1
  - Grade II or 2
  - Grade III or 3
  - 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"
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

HOSPITAL NAME/ADDRESS

PATIENT NAME/INFORMATION

(continued on next page)
Illustration
Indicate on diagram primary tumor and regional nodes involved.