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Retinoblastoma - Childhood - Stages [1]

This section has been reviewed and approved by the Cancer.Net Editorial Board [2], 04/2013

ON THIS PAGE: You will learn about how doctors describe a cancer?s growth or spread. This is called the stage. To see other pages, use the menu on the side of your screen.

Staging is a way of describing where the cancer is located, if or where it has spread, and whether it is affecting other parts of the body. Doctors use diagnostic tests to determine the cancer's stage, so staging may not be complete until all the tests are finished. Knowing the stage helps the doctor to decide what kind of treatment is best and can help predict a patient's prognosis (chance of recovery). There are different stage descriptions for different types of cancer.

After a retinoblastoma has been found, the doctor will determine the extent of disease in the eye and if the disease has spread outside the eye. This is called staging, and it helps doctors plan treatment. First, the doctor will determine if the retinoblastoma is intraocular or extraocular.

Intraocular. This means that cancer occurs in one or both eyes, but has not spread into surrounding tissues or other parts of the body.

Extraocular. The cancer has spread to tissues around the eye or to other parts of the body.

Several intraocular staging systems have been in use for many years to help doctors plan treatment. However, the recent success of clinical trials (research studies) demonstrating the effectiveness of chemotherapy to shrink tumors has led to the development of a newer staging system called the International Classification System.

International (ABC) Classification System

Group A:

- Small tumor(s) located only in the retina
- No tumor is larger than 3 millimeters (mm)
- No tumor is closer than 2 disc diameters (DD) from the fovea (the central ?pit? of the retina) or 1 DD from the optic nerve
- No vitreous seeding (tumor floating in the eye) or retinal detachment

Group B:

- Tumor(s) located only in the retina
- Any location in the retina
- No vitreous seeding
- No retinal detachment more than 5 mm from tumor base

Group C:

- Fine diffuse (spread throughout) or localized (located in one spot) vitreous seeding
- Retinal detachment: more than Group B and up to total retinal detachment
- No vitreous/sub-retinal ?snowballs? or masses

Group D:

- Massive vitreous/sub-retinal seeding
- Vitreous or sub-retinal snowballs/masses
- Retinal detachment: more than Group B and up to total retinal detachment

Group E:

No visual potential, **or**:

Presence of one or more:

- Tumor in CB/anterior segment
- Neovascular glaucoma (This usually results in increased pressure within the eye as a result of blood vessels growing into the anterior segment of the eye and represents an advanced stage of retinoblastoma.)
- Vitreous hemorrhage (bleeding from the eye)
- Phthisical/prephthisical (deteriorating) eye
- Hyphema (blood in the anterior of the eye)/corneal staining
- Orbital cellulitis-like presentation (the eye looks similar to the signs of an infection, but there is no infection)
- Tumor anterior to anterior hyaloids (the tumor is behind the thin membrane that separates the vitreous (white gel-like part of the eye) from other parts of the eye)

Extraocular staging

This staging system applies to cancer that has spread outside of the eye, either to the tissues around the eye or the central nervous system (CNS), bone marrow, or lymph nodes.

Stage 0 (zero). This means that the cancer is in the eye (called intraocular) and has not spread outside of the eye. Patients with stage 0 disease should be classified based on the International Classification System, outlined above. Doctors recommend conservative treatment for this stage with no additional adjuvant therapy (meaning no additional treatment after surgery; see the Treatment [3] section for additional information).

Stage I. A classification of stage I means that eye has already been removed and there is some microscopic spread to the optic nerve. These patients need adjuvant chemotherapy after surgery.

Stage II. This stage means that the tumor has spread to the optic nerve or the sclera (the white part of the eye). Patients with this stage need adjuvant chemotherapy and possibly radiation therapy (see the Treatment [3] section for more information on these treatments.)

Stage III. This means that the cancer has spread to the lymph nodes or the bony cavity that surrounds the eyeball. It is divided into two substages called stage IIIa and stage IIIb, depending on where the tumor has spread. The treatment for this stage involves both chemotherapy and radiation therapy.

Stage IV. In stage IV, the tumor has spread to distant areas of the body outside the eye, via the lymphatic system and blood vessels. Stage IV is divided into substages of IVa and IVb. Stage IVb is further divided into IVb1, IVb2, and IVb3, depending on the location of the spread. This stage of extraocular retinoblastoma is treated with high-dose chemotherapy followed by stem cell rescue. This means that the child?s stem cell/bone marrow cells are replaced with healthy cells. Learn more about bone marrow/stem cell transplantation [4]. Radiation therapy may also be used in addition to high-dose chemotherapy for some patients with stage IV disease.

Recurrent. Recurrent cancer is cancer that has come back after treatment. If there is a recurrence, the cancer may need to be staged again (called re-staging) using the system above.

Information about the cancer?s stage will help the doctor recommend a treatment plan. The next section helps explain the treatment options for this type of cancer. Use the menu on the side of your screen to select Treatment Options, or you can select another section, to continue reading this guide.

Links:
[1] http://www.cancer.net/cancer-types/retinoblastoma-childhood/stages
[2] http://www.cancer.net/about-us
[3] http://www.cancer.net/node/19583
[4] http://www.cancer.net/node/24409