

# Retinoblastoma for the Pediatric Patient

## What is retinoblastoma?

Retinoblastoma is a cancerous tumor of the retina. The retina is the thin nerve tissue in the back of the eye that senses light and forms images. Retinoblastoma may be hereditary or nonhereditary. The hereditary form may be in one or both eyes and generally affects younger children (median age is 7 months). Most retinoblastoma occurring in only one eye is not hereditary and is more often found in older children (median age is 23 months). Retinoblastoma is a rare disease but is the most common eye tumor in children. Children with a family history of retinoblastoma should have periodic examinations, including genetic counseling, to determine their risk for developing the disease.

Early detection and diagnosis are important because retinoblastoma grows very rapidly. The smaller the tumor is at diagnosis, the greater the likelihood that vision can be retained. Tumor spread at the time of diagnosis is rare but can occur when the tumor is large and has had the opportunity to grow along the eye nerve and back into the space in and around the brain. The tumor can also spread to the bone marrow, bone, lymph nodes, and other organs.

## What are the symptoms of retinoblastoma?

Leukocoria (“cat’s eye reflex”) is the most common presentation of retinoblastoma. Leukocoria occurs when reflected light from the tumor appears white through the pupil. Strabismus (wandering or “crossed” eye), decreased vision in one eye, painful eyes, and erythematous conjunctivae (“pinkeye”) are other symptoms.

## How is retinoblastoma diagnosed and treated?

If retinoblastoma is suspected, the child will need to have a careful eye exam under anesthesia. An MRI or CT scan of the eyes and brain is also obtained. If the tumor has spread beyond the globe of the eye, the doctor will need to obtain bone scans, a bone marrow aspiration, and a spinal tap to find out if the cancer has spread from the eye to other parts of the body. This is called staging. It is important to know the stage of the disease to plan treatment.

The type of treatment given depends on the extent of disease within the eye, whether the disease is in one or both eyes, and whether the disease has spread beyond the eye. Treatment options that attempt to cure and preserve or recover useful vision include the following:

- Enucleation – removal of the eye.
- Radiation therapy – use of high dose x-rays to kill cancer cells.
- Cryotherapy – use of extreme cold to destroy cancer cells.
- Photocoagulation – use of laser light to destroy blood vessels that supply blood to the tumor.
- Thermotherapy – use of heat to destroy cancer cells.
- Chemotherapy – use of drugs to kill cancer cells.