# Patient Education



Making Cancer History®

# **Retinoblastoma for the Pediatric Patient**

## What is retinoblastoma?

Retinoblastoma is a cancerous tumor of the retina (the back of the eye that senses light and forms images) and is the most common eye tumor in children. Retinoblastoma is most common in young children and is very rare in adolescents and adults. Retinoblastoma tumors can occur in one or both eyes and can rarely spread outside of the eyes to other parts of the body. Retinoblastoma tumors are associated with defects in the *Rb* gene, which controls how retina cells grow.

Retinoblastoma may be hereditary or non-hereditary. The hereditary form may be in one or both eyes and generally affects younger children (median age is 7 months). The non-hereditary form is more often found in older children (median age is 23 months) who have retinoblastoma in only one eye.

Early detection and diagnosis are important because retinoblastoma can grow very rapidly. The smaller the tumor is at diagnosis, the greater the likelihood that vision can be retained. Tumor spread is more likely to 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. Children with a family history of retinoblastoma should have periodic examinations, including genetic counseling, to determine their risk for developing the disease.

### 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 erythmatous conjunctivae ("pinkeye") are other symptoms.

### How is retinoblastoma diagnosed and treated?

If retinoblastoma is suspected, the child will need to have a careful eye examination, and may need to be asleep for a thorough examination. An MRI or CT scan of the eyes and brain may also be obtained. If the tumor has spread beyond the globe of the eye, the doctor may need to obtain bone scans, a bone marrow aspiration, or a spinal tap to find out if the cancer has spread to other parts of the body. 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.