

Treatment for Localized Retinoblastoma

Your child has recently been diagnosed with retinoblastoma. This is a cancer in one or both eyes. This is a very treatable type of cancer.

Treatment

Your child's doctor will discuss the treatment options with you. These options may include:

- **Lasers or cryotherapy (freezing):** This method is for treating small tumors in the eye.
- **Chemotherapy:** This uses anti-cancer drugs to shrink large tumors. This is followed by laser or cryotherapy to treat any tumor that is left.
- **Surgery:** When the tumors are large and there is no chance of saving vision, removing the eye is best.

Types of Chemotherapy (Chemo)

- **Intra-arterial:** This treatment infuses the chemotherapy directly into a blood vessel in the eye. This procedure is also called selective optic artery chemotherapy.
- **Systemic:** For very young patients or those with disease outside the eye, the chemotherapy is given into the blood stream through a port or catheter.

Intravenous Line (IV)

Your child will receive repeated eye exams under anesthesia (while asleep). This will require starting an intravenous line (a small plastic tube into a vein). If getting intra-arterial chemo only, they will not need any other IV line.

Port-a-Cath or Hickman Catheter

For systemic chemotherapy, your child will need a Port-a-Cath or Hickman catheter. This is a long lasting IV line. The Port-a-Cath is placed under the skin and the Hickman line comes out of the skin. Both have tubing inside the body that go into large vessels leading to the heart. With either of these we can draw blood, give chemo and manage any problems of treatments. The surgical placement of this line will be done before starting treatment.

Intra-arterial Chemotherapy

This treatment is for children over 6 months old. This is done by interventional radiologists. They pass a catheter from your child's groin into the arteries of the head and the ophthalmic artery, the artery going to the affected eye. This provides a large dose of chemo to the tumor but a small amount to the rest of the body.

During the treatment, your child will need to be asleep, and the doctor will use x-ray pictures to make sure the catheter is in the right place. Anti-clotting medicine is given to prevent blood clots. The procedure often takes 2 to 4 hours. At the end of the procedure the catheter is removed.

Your child will need to stay at the hospital for several hours after for close monitoring. Most children go home the same day, but some may have to stay overnight.

This treatment is given every 4 weeks for three or more times depending upon response of the tumor(s) in the eye.

Systemic Chemotherapy

This treatment is for patients under 6 months old or for patients with more severe eye disease.

During the **induction phase** (first phase) of treatment, your child will receive 4 to 6 treatment sessions in the infusion center. Each treatment session will last 2 days and follow the schedule below.

Day 1:

- **Vincristine** – given as a rapid infusion through the catheter.
- **Carboplatin** – given as a 60 to 90-minute infusion through the catheter.
- **Etoposide** – given as a one-hour infusion through the catheter.

Day 2:

- **Etoposide** – A second dose is given.

Your child will also receive fluids with treatment and be closely watched for any side effects and to see how the eye responds to treatment.

Eye Exams

Your child will be seen by an eye doctor for an eye exam (often under anesthesia). This is done about 4 weeks after the second course and every 4 weeks through later courses of treatment. The eye doctor may use treatment with laser or cryotherapy (see below) to kill small tumors at the eye exams even when your child is getting chemo.

These eye exams are done before the first day of the third two-day cycle. This will be done every four weeks until a minimum of four courses of chemo are given or until your child's eye tumors have been completely controlled. Six to eight courses may be needed, but rare. Additional eye exams and local treatment may be needed after completing the course.

Laser or Cryotherapy

When your child is examined under anesthesia (while asleep) by the eye doctor, small tumors in the eye(s) may be treated with light (laser) that is shined on the tumor(s) in the back half of the eye.

In some cases, the tumors may be in the front part of the eye and be treated with freezing and thawing, a technique called cryotherapy.

These local treatments to the tumor(s) are also used to completely kill the cancer cells in the tumor(s). These are not used to treat the larger tumors in the eye(s) seen in most children with retinoblastoma at diagnosis.

Most children are treated with chemotherapy first to try and shrink the tumors. When larger tumor(s) respond well to the treatment and the tumor(s) shrink in size, the use of laser or cryotherapy is the way to destroy the last parts of the cancer. The doctor will discuss the effects of local cryotherapy or laser. These are usually mild and consist of redness of the eye that lasts a day or two. Sometimes there can be permanent scarring, which can affect vision.

Continued Therapy

Complete control of the eye tumor(s) is when no growth or new tumors are found after two eye exams at least 4 weeks apart.

If some part of the remaining tumor(s) grow again, they can usually be treated with laser or cryotherapy. Some children have multiple tumors regrow and may need more chemo. Patients **will not** receive more than eight courses. For the patients who received chemo into the vein and have tumors that do not respond, they will change to intra-arterial chemotherapy.

Long-term Monitoring with Eye Exams

All patients will have eye exams under anesthesia on a frequent basis after treatment is done. These exams will be every four to eight weeks until there is no growth of old tumor(s) and no new tumor(s) over two exams. They will then be every three months until at least 18 months from the last active disease. After that they will be every 6 months for 18 months.

Genetic Testing

Genetic testing helps decide how long follow-up exams will be done. All patients have a defect in their genes in the tumor in the eye(s). For patients who **do not have** a defect in the genes in their blood, scheduled follow-up stops at 3 years from last active disease. For patients who **have** the gene defect in their blood, scheduled follow up continues once a year to 5 years from last active disease.

The use of anesthesia for eye exams stops when a child can be still so the doctor can use bright light to see all parts of the affected eye(s). Few children can sit still enough to do awake exams until about 5 years old. The above is a general schedule that will be custom-made for your child.

Who to Call

American Family Children's Hospital
Department of Pediatric
Hematology, Oncology and
Stem Cell Transplant

Clinic Phone: **(608) 263-6420**

8am - 5pm – Monday - Friday

Medical Related Calls:

(608) 262-0486

8am - 5pm – Monday - Friday

Ask for the Pediatric
Hematology/Oncology Nurse
Practitioner.

After hours, weekends, and holidays, ask for the Pediatric Hematology/Oncologist on-call.

References:

Ancona-Lezama D, Dalvin LA, Shields CL. Modern treatment of retinoblastoma: A 2020 review. *Indian J Ophthalmol.* 2020;68(11):2356-2365. doi:10.4103/ijo.IJO_721_20

Your health care team may have given you this information as part of your care. If so, please use it and call if you have any questions. If this information was not given to you as part of your care, please check with your doctor. This is not medical advice. This is not to be used for diagnosis or treatment of any medical condition. Because each person's health needs are different, you should talk with your doctor or others on your health care team when using this information. If you have an emergency, please call 911. Copyright © 8/2022 University of Wisconsin Hospitals and Clinics Authority. All rights reserved. Produced by the Department of Nursing. HF#8257.