

Eye Cancer Early Detection, Diagnosis, and Staging

Learn about the signs and symptoms of ocular melanoma. Find out how eye cancer is tested for, diagnosed, and staged.

Detection and Diagnosis

Catching cancer early often allows for more treatment options. Some early cancers may have signs and symptoms that can be noticed, but that is not always the case.

- Can Eye Cancer Be Found Early?
- Signs and Symptoms of Eye Cancer
- Tests for Eye Cancer

Stages and Outlook (Prognosis)

After a cancer diagnosis, staging provides important information about the extent of cancer in the body and anticipated response to treatment.

- Eye Cancer Stages
- Eye Cancer Survival Rates

Questions to Ask About Eye Cancer

Here are some questions you can ask your cancer care team to help you better understand your cancer diagnosis and treatment options.

Questions to Ask About Eye Cancer

Can Eye Cancer Be Found Early?

Eye cancer is uncommon, and there are no widely recommended screening tests for this cancer in people at average risk. (Screening is testing for a disease like cancer in people without any symptoms.) Still, some eye cancers can be found early.

Some doctors may recommend yearly eye exams for those at <u>higher risk of eye</u> <u>melanoma</u>¹, such as people with dysplastic nevus syndrome or BAP1 cancer syndrome . Regular eye exams are an important part of everyone's health care, even if they have no symptoms. Often melanomas of the eye are found during a routine eye exam. When the doctor looks through the pupil at the back of the eye, they may see a dark spot that might be an early melanoma.

Many doctors feel that most melanomas start from a *nevus* (mole), which is a benign (non-cancerous) tumor of pigment cells. If an eye nevus is present, it should be looked

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Signs and Symptoms of Eye Cancer

Many people with eye melanoma don't have symptoms unless the cancer grows in certain parts of the eye or becomes more advanced. Other, less serious conditions can also cause many of these symptoms. For example, floaters can be a normal part of the aging process. Still, if you have any of these symptoms, it's important to see a doctor right away so the cause can be found and treated, if needed.

Signs and symptoms of eye melanomas can include:

- Problems with vision (blurry vision or sudden loss of vision)
- Floaters (spots or squiggles drifting in the field of vision) or flashes of light
- Visual field loss (losing part of your field of sight)
- A growing dark spot on the colored part of the eye (iris)
- Change in the size or shape of the pupil (the dark spot in the center of the eye)
- Change in position of the eyeball within its socket
- Bulging of the eye
- Change in the way the eye moves within the socket

Pain is rare unless the tumor has grown extensively outside the eye.

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Tests for Eye Cancer

- Eye exam
- Imaging tests
- Biopsy
- Blood tests

Eye exam

Examination of the eye by an ophthalmologist (a medical doctor specializing in eye diseases) is often the most important first step in diagnosing melanoma of the eye. The doctor will ask if you are having any symptoms and check your vision and eye movement. The doctor will also look for enlarged blood vessels on the outside of the eye, which can be a sign of a tumor inside the eye.

The ophthalmologist may also use special instruments to get a good look inside the eye for a tumor or other abnormality. You may get drops in your eye to dilate the pupil before the doctor uses these instruments.

- An **ophthalmoscope** (also known as a *direct ophthalmoscope*) is a hand-held instrument consisting of a light and a small magnifying lens.
- To get a more detailed view than with a direct ophthalmoscope, an **indirect ophthalmoscope** or a **slit lamp** may be used. With either instrument, the doctor looks into your eye through a stronger magnified lens, but the slit lamp tends to have more magnification and sits still on a platform in front of you. With an indirect ophthalmoscope, the doctor has you recline a bit, opens your eye, and holds the

magnifying lens very close to it while a bright light shines into the eye.

• A **gonioscopy lens** is a specially mirrored lens that is placed on the cornea (the outer part of the eye) after it is numbed. It can be used to look for tumor growth into areas of the eye that would otherwise be hard to see.

Even if you recently had an eye exam, if you start to have any symptoms, get another exam. Sometimes these tumors are missed or grow so fast that they weren't there when you were last examined.

If an eye exam suggests you might have eye cancer, more tests such as imaging tests or other procedures might be done to confirm the diagnosis.

Imaging tests

Imaging tests use sound waves, x-rays, or magnetic fields, or radioactive particles to create pictures of the inside of your body. These tests might be done for a number of reasons, including:

- To help find a suspicious area that might be cancer
- To help determine the stage(extent) of the cancer
- To help show if treatment is working
- To look for possible signs of <u>cancer coming back</u>¹after treatment

Ultrasound

<u>Ultrasound</u>² is a very common test used to help diagnose eye melanomas. Ultrasound uses sound waves and their echoes to make pictures of internal organs or masses. For this test, a small wand-like instrument is placed up against the eyelid or eyeball, sends sound waves through the eye, and picks up the echoes as they bounce off the organs. The echoes are converted into an image on a computer screen.

This test is especially useful for diagnosing eye melanomas because they look a certain way on ultrasound. Using this test, doctors can confirm a diagnosis of melanoma of the eye in most cases. This test can also show the location and the size of the tumor. If you have already been diagnosed with eye melanoma, an ultrasound of your abdomen may be done to look for tumors in the liver, which is a common site of spread of this cancer.

Ultrasound biomicroscopy (UBM): This is a special type of ultrasound that uses highenergy sound waves to create very detailed images of the front parts of the eye.

Optical coherence tomography (OCT)

This test is similar to an ultrasound, but it uses light waves instead of sound waves to create very detailed images of the back of the eye.

Fluorescein angiography

For this test, an orange fluorescent dye (fluorescein) is injected into the bloodstream through a vein in the arm. Pictures of the back of the eye are then taken using a special light that makes the dye fluoresce (glow). This lets the doctor see the blood vessels inside the eye. Although melanomas don't have a special appearance with this test, some other eye problems do. Doctors can use this method to tell if something is not a melanoma.

Chest x-ray

If you have been diagnosed with eye melanoma, an <u>x-ray of your chest³</u> may be done to see if the cancer has spread to your lungs.

Computed tomography (CT) scan

A <u>CT scan</u>⁴ combines many x-rays to make detailed cross-sectional images of parts of the body. This scan is sometimes used to see if a melanoma has spread outside of the eye into nearby structures. It may also be used to look for spread of the cancer to distant organs such as the liver.

Magnetic resonance imaging (MRI) scan

<u>MRI scans</u>⁵ are particularly useful for looking at eye tumors and spread of tumor outside the eye orbit in places like the liver. MRIs provide detailed images of soft tissues in the body, but use radio waves and strong magnets instead of x-rays.

Biopsy

For most types of cancer, the diagnosis is made by removing a small piece of the tumor and looking at it in the lab for cancer cells. This is known as a <u>biopsy⁶</u>.

A biopsy is often not needed to diagnose eye melanomas because almost all cases can be accurately diagnosed by the eye exam and imaging tests. Sometimes, a biopsy may be useful to check for certain gene mutations (changes) that can predict outcomes (prognosis) as well as help choose targeted drugs for your cancer. Also, certain eye melanomas can spread for many years before they are diagnosed so doing a biopsy of a worrisome area early may be helpful.

If a biopsy is needed, it can be done either with sedation and local anesthesia (numbing medicine) or while a person is under general anesthesia (in a deep sleep). Different types of biopsies can be done for eye melanoma depending on where it is located including:

- A <u>FNA</u>⁷ (fine needle aspiration): Using a thin needle to remove a small sample of aqueous humor (the liquid between the cornea and the lens)
- An incisional or excisional biopsy⁸ (cutting out either part of or all of the tumor)
- A fine needle biopsy of the tumor: Cells from the tumor are sucked up into a syringe through a small needle and examined in the lab.

Newer techniques help to lower the chances of tumor cells leaking and spreading along the needle path during these biopsies so the cancer doesn't spread within or outside the eye.

While most people with melanoma of the eye are treated without having a biopsy first, your doctor may recommend a biopsy depending on your specific situation. They can discuss the risks and benefits of the procedure they feel is best for you. Some doctors have started using biopsies to get a sample of the tumor for gene testing (DecisionDx-UM). They have found that certain patterns of genes in tumor cells are a good way to tell if an eye melanoma is likely to spread. Based on these gene patterns, a little more than half of eye melanomas are shown to be Class 1 (1A or 1B) tumors which have a low risk of spreading. The remaining eye melanomas fall into the Class 2 category, which have a very high risk of spreading. See <u>What's New in Eye Cancer Research?</u>⁹ for more information.

Liquid biopsy

A new type of biopsy called a liquid biopsy is being looked at more often. Instead of having to make a cut or put a needle into the eye, melanoma tumor cells can be collected from a blood sample. These cancer cells can then be tested for certain traits, including genetic changes, that can help predict how likely the cancer is to spread or come back after treatment. Liquid biopsies might help diagnose tumor spread earlier, or help the doctors know if treatment is working. This could be very helpful in people who did not have a biopsy of the tumor and want to preserve their vision. However, the equipment needed for this test is not readily available so this type of biopsy is not done routinely and is mainly done as part of a clinical trial.

Blood tests

Blood tests can't be used to diagnose melanoma of the eye, but they may be done once a diagnosis is made.

Liver function tests

If you have been diagnosed with eye melanoma, your doctor may order blood tests to see how well your liver is working. Abnormal test results can sometimes be a sign that the cancer has spread to the liver.

Hyperlinks

- 1. www.cancer.org/cancer/survivorship/long-term-health-concerns/recurrence.html
- 2. <u>www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/ultrasound-for-</u> <u>cancer.html</u>
- 3. <u>www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/x-rays-and-other-radiographic-tests.html</u>
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- 5. www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/mri-for-cancer.html
- 6. <u>www.cancer.org/cancer/diagnosis-staging/tests/biopsy-and-cytology-tests.html</u>
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- 9. <u>www.cancer.org/cancer/types/eye-cancer/about/new-research.html</u>

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Eye Cancer Stages

- How is the stage determined?
- AJCC TNM staging system for melanoma of the eye
- Collaborative Ocular Melanoma Study (COMS) staging of melanoma of the eye

After someone is diagnosed with eye cancer, doctors will try to figure out if it has spread, and if so, how far. This process is called **staging**. The stage of a cancer describes how much cancer is in the body. It helps determine how serious the cancer is and how best to <u>treat</u>¹ it. Doctors also use a cancer's stage when talking about survival statistics.

How is the stage determined?

The cancer stage is determined from the results of eye exams, imaging tests (ultrasound, CT or MRI scan, etc.) and other tests. (See Tests for Eye Cancer.)

A staging system is a standard way for the cancer care team to describe how far a cancer has spread. The most common systems used to describe the stages of eye melanomas are the American Joint Committee on Cancer (AJCC) TNM system and the system used by the Collaborative Ocular Melanoma Study (COMS) group.

AJCC TNM staging system for melanoma of the eye

Most eye melanomas start in the uvea, which includes the iris, ciliary body, and choroid. (See <u>What Is Eye Cancer</u>²?) The system below is for these **uveal melanomas**.

Less often, melanomas can start in other areas in or around the eye, some of which have their own staging systems (such as conjunctival melanoma). Talk to your doctor to learn more about your stage if you have a less common type of eye melanoma.

The system described below is the most recent AJCC system, effective January 2018.

The TNM system is based on 3 key pieces of information:

- The size and extent of the main **tumor (T)**: How large is the eye tumor? Has it invaded into nearby structures?
- The spread to nearby lymph **nodes (N):** Has the cancer spread to the nearby lymph nodes around the ear or neck? Has the cancer spread to (not grown into) other parts of the eye?
- The spread (**metastasis**) to distant sites **(M)**: Has the cancer spread to distant parts of the body? (The most common site of spread is the liver.)

Numbers or letters appear after T, N, and M to provide more details about each of these factors. Higher numbers or letters mean the cancer is more advanced.

The T categories for iris melanomas are different from the T categories for ciliary body and choroidal melanomas. But the N and M categories are the same for melanomas in all 3 parts of the uvea.

T categories for iris melanoma

TX: The primary tumor cannot be assessed; information not known.

T0: No evidence of a primary tumor.

T1: Tumor is only in the iris.

- **T1a:** The tumor is only in the iris and touches 1/4 or less of the iris.
- **T1b:** The tumor is only in the iris and touches more than 1/4 of the iris.
- **T1c:** The tumor is only in the iris and is causing an increase in the eye pressure (glaucoma).

T2: Tumor has grown into the ciliary body or choroid (or both).

- **T2a:** Tumor has grown into the ciliary body.
- **T2b:** Tumor has grown into the ciliary body and choroid.
- **T2c:** Tumor has grown into the ciliary body, choroid, or both, and it is causing glaucoma.
- **T3:** Tumor has grown into the ciliary body and/or choroid and into the sclera.

T4:

inch) or less across.

T2 tumors:

T4e: The tumor can be any size. It is growing outside the eyeball and the part of the tumor that is outside the eyeball is greater than 5 mm across.

N categories for iris, ciliary body, and choroidal melanomas

NX: Lymph nodes cannot be assessed.

N0: Cancer has not spread to nearby lymph nodes.

N1: Cancer has spread to nearby lymph nodes, or it has spread as small cancer deposits in other parts of the eye.

- N1a: Cancer has spread to nearby lymph nodes.
- **N1b:** Cancer has not spread to nearby lymph nodes, but it has spread as small cancer deposits in other parts of the eye.

M categories for iris, ciliary body, and choroidal melanomas

M0: Cancer has not spread to distant parts of the body.

M1: Cancer has spread to distant parts of the body.

- M1a: The largest area of cancer spread is no more than 3 centimeters (cm) a little over an inch across.
- **M1b:** The largest area of cancer spread is between 3.1 and 8 cm across (8 cm is a little over 3 inches).
- M1c: The largest area of cancer spread is 8.1 cm or more across.

Stage grouping

To assign an overall stage, the T, N, and M categories are combined in a process called **stage grouping**. The stages are described by Roman numerals from I (the least advanced) to IV (the most advanced). Some stages are further divided with letters.

Stage TNM categories

Stage I T1a, N0, M0

Stage IIA T1b to T1d, N0, M0 OR T2a, N0, M0

Stage IIB T2b or T3a, N0, M0

Stage IIIA T2c or T2d, N0, M0 OR T3b or T3c, N0, M0 OR T4a, N0, M0

Stage IIIB T3d, N0, M0 **OR** T4b or T4c, N0, M0

Stage IIIC T4d or T4e, N0, M0

Stage IV Any T, N1, M0 OR Any T, any N, M1

This staging system for uveal melanoma can be very complex. If you're interested in learning more about it and how it might apply to your cancer, ask your doctor to explain it to you in a way you understand.

Collaborative Ocular Melanoma Study (COMS) staging of melanoma of the eye

The TNM system is very detailed, but in practice doctors may use the simpler staging system devised by the COMS group, which has done most of the clinical research on how to treat intraocular melanoma. This system divides eye melanomas into small, medium, and large:

- Small: Between 1 mm and 3 mm in height and between 5 mm and 16 mm across
- Medium: Between 3.1 mm and 8 mm in height and no more than 16 mm across
- Large: More than 8 mm in height or more than 16 mm across

Hyperlinks

- 1. www.cancer.org/cancer/types/eye-cancer/treating.html
- 2. www.cancer.org/cancer/types/eye-cancer/about/what-is-eye-cancer.html

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Eye Cancer Survival Rates

- What is a 5-year relative survival rate?
- Where do these numbers come from?
- 5-year relative survival rates for eye melanoma
- Understanding the numbers

Survival rates can give you an idea of what percentage of people with the same type and stage of cancer are still alive a certain amount of time (usually 5 years) after they were diagnosed. They can't tell you how long you will live, but they may help give you a better understanding of how likely it is that your treatment will be successful.

Keep in mind that survival rates are estimates and are often based on previous

The SEER database tracks 5-year relative survival rates for eye cancer (ocular melanoma) in the United States, based on how far the cancer has spread. The SEER database, however, does not group cancers by AJCC TNM stages (stage 1, stage 2, stage 3, etc.). Instead, it groups cancers into localized, regional, and distant stages:

- Localized: There is no sign that the cancer has spread outside of the eye.
- **Regional:** The cancer has spread outside the eye to nearby structures or lymph nodes.
- Distant: The cancer has spread to distant parts of the body, such as the liver.

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These numbers are based on people diagnosed with melanoma of the eye (also known as ocular melanoma or intraocular melanoma) between 2012 and 2018.

SEER* stage	5-year relative survival rate0 0 1 72 566.5	52 Tm /F2 172
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Hyperlinks

1. www.cancer.org/cancer/types/eye-cancer/about/what-is-eye-cancer.html

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Questions to Ask About Eye Cancer

- When you're told you have eye cancer
- When deciding on a treatment plan
- During treatment
- After treatment

It's important to have honest, open discussions with your cancer team. They want to answer all your questions, so that you can make informed treatment and life decisions. For instance, consider these questions:

When you're told you have eye cancer

- What kind of eye cancer¹ do I have?
- Has my cancer spread beyond the eye?
- What is the stage (extent) of my cancer, and what does that mean?
- Will I need any other tests before we can decide on treatment?
- Will I need to see any other types of doctors?

- Are there other factors that could affect my treatment options?
- If I'm concerned about the costs and insurance coverage for my diagnosis and treatment, who can help me?

When deciding on a treatment plan

- How much experience do you have treating this type of cancer?
- Should I get a second opinion? Can you recommend a doctor or cancer center?

• What symptoms should I watch for?

How will we know if the eye cancer has come back? What would my options be if

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