

About Eye Cancer

Get an overview of eye cancer (ocular melanoma) and the latest key statistics in the US.

Overview and Types

If you have been diagnosed with eye cancer or are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

• What Is Eye Cancer?

Research and Statistics

See the latest estimates for new cases of eye cancer and deaths in the US and what research is currently being done.

- Key Statistics for Eye Cancer
- What's New in Eye Cancer Research?

What Is Eye Cancer?

- Where eye cancers start
- Cancers in the eye (intraocular cancers)
- Orbital and adnexal cancers

Eye cancer can refer to any cancer that starts in the eye. Cancer starts when cells begin to grow out of control. (To learn about how cancers start and spread, see <u>What Is</u> <u>Cancer?</u>¹)

The most common type of eye cancer is melanoma. But there are other types of cancer that affect different kinds of cells in the eye.

Where eye cancers start

The eye has 3 major parts:

- The **eyeball** (globe), which is mostly filled with a jelly-like material called vitreous humor and has 3 main layers (the sclera, the uvea, and the retina)
- The **orbit** (the tissues surrounding the eyeball)
- The adnexal (accessory) structures such as the eyelids and tear glands

Different types of cancer start in each of these areas.

Cancers in the eye (intraocular cancers)

Cancers that are within the eye itself are called intraocular cancers:

- Primary intraocular cancers start in the eye.
- Secondary intraocular cancers start in another part of the body and spread to the eye.

In adults, the most common primary intraocular cancers are:

- Melanoma (Intraocular melanoma is the focus of our information on eye cancer.)
- Non-Hodgkin lymphoma (See <u>Non-Hodgkin Lymphoma (NHL)</u>² for more information on primary intraocular lymphoma.)

In children, the most common primary intraocular cancers are:

- **Retinoblastoma**, a cancer that starts in cells in the retina (the light-sensing cells in the back of the eye)
- Medulloepithelioma, the second most common type, which is still extremely rare.

These childhood cancers are discussed in <u>Retinoblastoma³</u>.

Secondary intraocular cancers (cancers that start somewhere else in the body and then spread to the eye) are not truly "eye cancers," but they are actually more common than primary intraocular cancers. The most common cancers that spread to the eye are <u>breast</u>⁴ and <u>lung cancers</u>⁵. Most often these cancers spread to the part of the eyeball called the *uvea*.

Intraocular melanoma (melanoma of the eye)

Intraocular melanoma is the most common type of cancer that develops within the eyeball in adults, but it is still fairly rare. Melanomas that start in the skin are much more common than melanomas that start in the eye. Melanomas develop from pigment-making cells called *melanocytes*. When melanoma develops in the eye, it is usually in the uvea (**uveal melanoma**) and rarely in the conjunctiva (**conjunctival melanoma**).

Uveal melanomas

The uvea is the middle layer of the eyeball. It has 3 main parts:

- The **iris** is the colored part of the eye (most often blue or brown). It surrounds the pupil, the small opening where light enters the eyeball.
- The **choroid** is a thin, pigmented layer lining the eyeball that nourishes the retina and the front of the eye with blood.
- The **ciliary body**contains the muscles inside the eye that change the shape of the lens so that the eye can focus on near or distant objects. It also has cells that make aqueous humor, the clear fluid in the front of the eye between the cornea and the lens.

About 9 out of 10 intraocular melanomas develop in the choroid or ciliary body. Choroid cells make the same kind of pigment as melanocytes in the skin, so it's not surprising that these cells sometimes form melanomas.

Most of the other intraocular melanomas start in the iris. These are the easiest for a person (or their doctor) to see because they often start in a dark spot on the iris that has been present for many years and then begins to grow. These melanomas usually are slow growing, and they rarely spread to other parts of the body. For these reasons, people with iris melanomas generally have a good prognosis (outlook).

Uveal melanomas can spread through the blood, most often to the liver.

Conjunctival melanomas

The conjunctiva is a thin clear covering over the sclera. (The sclera is the tough, white covering over most of the outside of the eyeball. In the front of the eye it is continuous with the cornea, which is clear to let light through.)

Conjunctival melanomas are extremely rare. They tend to be more aggressive and grow into nearby structures. Because they can spread through the blood and the lymph system, they can also spread to distant organs like the lungs, liver, or brain, where the cancer can become life-threatening.

Orbital and adnexal cancers

The **orbit** consists of the tissues surrounding the eyeball. These include muscles that move the eyeball in different directions and the nerves attached to the eye. Cancers of these tissues are called *orbital cancers*.

Adnexal (accessory) structures include the eyelids and tear glands. Cancers that develop in these tissues are called *adnexal cancers*.

Cancers of the orbit and adnexa develop from tissues such as muscle, nerve, and skin around the eyeball and are like cancers in other parts of the body. For example:

- Cancers of the eyelid are usually skin cancers. (See <u>Melanoma Skin Cancer⁶</u> or <u>Skin Cancer: Basal and Squamous Cell⁷.)</u>
- For cancer affecting the eye muscles, see <u>Rhabdomyosarcoma</u>⁸.

Hyperlinks

- 1. www.cancer.org/cancer/understanding-cancer/what-is-cancer.html
- 2. www.cancer.org/cancer/understanding-cancer/what-is-cancer.html
- 3. www.cancer.org/cancer/types/non-hodgkin-lymphoma.html
- 4. <u>www.cancer.org/cancer/types/retinoblastoma.html</u>
- 5. <u>www.cancer.org/cancer/types/breast-cancer.html</u>
- 6. www.cancer.org/cancer/types/lung-cancer.html
- 7. www.cancer.org/cancer/types/melanoma-skin-cancer.html
- 8. www.cancer.org/cancer/types/basal-and-squamous-cell-skin-cancer.html
- 9. www.cancer.org/cancer/types/rhabdomyosarcoma.html

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Key Statistics for Eye Cancer

The American Cancer Society's estimates for eye cancer in the United States for 2024 are:

- About 3,320 new cancers (mainly melanomas) of the eye and orbit (1,780 in men and 1,540 in women)
- About 560 deaths from cancers of the eye and orbit (260 in men and 300 in women)

Primary eye cancers can occur at any age, but the risk for most types increases as people get older. The rate of uveal melanomas has been fairly stable over the past few decades, but the rate of conjunctival melanomas has increased. Cancers that spread to the eye from another part of the body (secondary eye cancers) are actually more common than primary eye cancers.

Most cancers of the eye and orbit in adults are melanomas, but this cancer starts more often in other parts of the body. More than 9 out of 10 melanomas start in the skin.

Melanoma of the eye is much more common in White than in Black people, and is slightly more common in men than women.

For statistics on survival, see Eye Cancer Survival Rates¹.

Visit the <u>American Cancer Society's Cancer Statistics Center²</u> for more key statistics.

Hyperlinks

- 1. <u>www.cancer.org/cancer/types/eye-cancer/detection-diagnosis-staging/survival-</u> <u>rates.html</u>
- 2. cancerstatisticscenter.cancer.org/

References

American Cancer Society. *Cancer Facts & Figures 2024.* Atlanta, Ga: American Cancer Society; 2024.

Last Revised: January 17, 2024

What's New in Eye Cancer Research?

- Genetics
- New tests for eye cancer
- Advances in treatment

Many medical centers around the world are doing research on the causes and treatment of eye cancers. These are challenging diseases to study because they are not common. But each year scientists find out more about what causes them and how to improve treatment.

Genetics

Learning more about the gene changes that make eye cancer cells different from normal cells will likely play an important role in treating eye melanomas in the future.

Using genes to help find people at higher risk

Recently, researchers have found that patterns of gene expression in tumor cells appear to be an even better 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" tumors. These cancers have a low risk of spreading. The remaining eye melanomas fall into the "Class 2" category, which have a very high risk of spreading.

Some doctors now offer a test (DecisionDx-UM) for these gene changes, and some patients may want to have them to learn if their cancer is likely to spread. If a patient is found to be at high risk, the doctor might follow them more closely to try to detect cancer spread as early as possible. But other doctors are not as keen on using the test at this time, because we don't yet have proven ways to prevent the cancer spread or alter the outcome in people who are in the high risk group.

Using genes to help find new treatments

Identifying gene changes in eye cancer cells might also provide specific targets for newer drugs. For example, most eye melanomas have changes in either of 2 related genes, *GNAQ* or *GNA11*. The proteins made by these genes are part of the *MAPK* signaling pathway inside cells that helps them grow. It's not yet clear if drugs will be able to target these proteins directly, but drugs that target other proteins in the *MAPK* pathway are now being studied for use against eye melanomas, and some have shown early promising results. (See "Targeted therapy" below.)

New tests for eye cancer

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.

Advances in treatment

Immunotherapy

Immunotherapies are treatments that boost the body's immune system to help it attack

the cancer. Cytokines, monoclonal antibodies, cancer vaccines, and other immunotherapies are among the most promising approaches for treating melanoma. Although most clinical trials of these treatments focus on people with melanomas of the skin, results of these studies might help treat people with eye melanomas as well. For example:

- Drugs known as **immune checkpoint inhibitors**, which help boost the body's immune response, have been shown to be helpful in many people with skin melanomas. They might also be an option in some people with eye melanomas.
- **Bispecific T cell engagers (BiTEs)** are drugs that help bring immune cells called T cells together with melanoma cells, which can help boost the body's immune response. One of these drugs, tebentafusp, is now an option to treat some people with advanced eye melanomas.

Other new immunotherapy drugs are now being studied as well.

Targeted therapy drugs

As researchers have learned more about some of the changes in cells that cause them

now being studied for use against melanoma of the eye as well, including sunitinib, sorafenib, vorinostat, and everolimus.

To learn more about immunotherapy and targeted drugs now being used to treat eye melanoma, see <u>Targeted Drugs and Immunotherapy for Eye Cancer</u>².

Hyperlinks

- 1. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-</u> <u>trials.html</u>
- 2. www.cancer.org/cancer/types/eye-cancer/treating/targeted-therapy.html

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