**OPHTHALMOLOGY (EYE AND VISION)**

**OPHTHALMOLOGY**

**DEFINITION/DESCRIPTION**

Ophthalmology is the branch of medicine that focuses on the health of the eyes, including the diagnosis, treatment, and prevention of various eye diseases and conditions that may affect the eye and visual system. The term "ophthalmology" comes from the ancient Greek words "ophthalmos," meaning eye, and "logia," meaning study or discourse. This field encompasses a wide range of medical practices aimed at preserving eye health and treating various eye conditions. Ophthalmologists are medical doctors who specialize in diagnosing and treating eye disorders, and they undergo extensive training that includes both medical and surgical aspects of eye care

The term ophthalmology comes from the Greek roots ὀφθαλμός (ophthalmos, "eye") and -λογία (-logia, "study, discourse"), meaning "the study of eyes".

Ophthalmologists can diagnose and treat various eye diseases and conditions, including cataracts, retinal diseases, glaucoma, corneal diseases, eyelid and orbital disorders, uveitis, strabismus, ocular neoplasms, and neuro-ophthalmologic disorders.

They use various diagnostic tests such as ophthalmoscopy, visual field tests, and optical coherence tomography to assess and treat eye conditions.

Ophthalmology has a long history, with significant contributions from ancient civilizations like the Egyptians, Greeks, and Romans, and modern advancements in technology and medical knowledge.

In summary, ophthalmology addresses a broad spectrum of eye diseases such as cataracts, glaucoma, diabetic retinopathy, macular degeneration, amblyopia, strabismus, retinal disorders, uveitis, keratoconus, and many others affecting vision and eye health.

These conditions can be managed with proper diagnosis and treatment, often including medications, surgery, or corrective lenses. Regular eye exams are important for early detection and management of these conditions.

**LIST OF DISEASES THAT MAY AFFECT THE EYE AND VISUALS**

The diseases in this diverse list include:

* Achromatopsia,
* Adie’s pupil
* Age-Related Macular Degeneration (AMD),
* Albinism
* Alzheimer's Disease, Dementia and the eye
* Amblyopia (Lazy Eye)
* Aniridia
* Anisocoria
* Anophthalmia and Microphthalmia
* Astigmatism
* Behçet's disease
* Bell's Palsy
* Best disease (best vitelliform macular dystrophy)
* Birdshot chorioretinopathy
* Blepharitis
* Blocked tear ducts
* Cataract
* Central Serous Retinopathy
* Charles Bonnet syndrome
* Chalazion
* Choroideremia
* Coats disease
* Coloboma
* Color Blindness
* Corneal Disease and Dystrophies / Corneal Ulcer
* Conjunctivitis (Pink Eye)
* Cytomegalovirus (CMV) retinitis
* Diabetic Retinopathy
* Dry Eye syndrome
* Duane Syndrome
* Ectropion
* Entropion
* Excessive tearing
* Endophthalmitis
* Eye melanoma
* Floaters and Flashes
* Fuch’s dystrophy
* Glaucoma
* Hemianopia
* Herpes Simplex Infection (Ocular)
* Horner Syndrome
* Hordeolum
* Keratitis
* Keratoconus
* Low Vision
* **Leber Congenital amaurosis**
* Leber hereditary optic neuropathy
* Macular Degeneration (Edema, Hole, and Pucker)
* Macular telangiectasia
* Meesmann corneal dystrophy
* Metabolic and Toxic Retinopathies (e.g., Hydroxychloroquine Retinal Toxicity)
* Myopia (Nearsightedness)
* Mydriasis
* Nasolacrimal duct obstruction
* Nystagmus
* Ocular herpes
* **Ocular mucous membrane pemphigoid**
* Ocular Tumors (e.g., Cancer of the Eye)
* Optic atrophy
* Orbital Fractures
* Papilledema
* Pelvic Inflammatory Disease (with ocular manifestations)
* Proptosis (bulging eyelids)
* Prostatitis (with ocular symptoms in some systemic diseases)
* Proliferative vitreoretinopathy
* Ptosis (drooping eyelid)
* Pterygium
* Refractive Errors (Myopia, Hyperopia, Presbyopia, Astigmatism)
* Retinal Detachment
* Retinitis Pigmentosa
* Retinopathy of Prematurity (ROP)
* Rious Retinal Vascular Occlusions (Retinal Vein Occlusions)
* Scleritis and Episcleritis
* Stargardt disease
* Stye (Hordeolum)
* Strabismus
* **Thyroid eye disease** (Grave's Eye disease)
* Trachoma
* Uveal melanoma
* Uveitis
* Vaginitis (ocular symptoms in systemic infections)
* Vitreous Hemorrhage
* Vitreomacular traction syndrome

**ACANTHAMOEBA KERATITIS**

*ALTERNATIVE NAMES:* Acanthamoeba keratitis is sometimes referred to as “amoebic keratitis”, or by similar names.

**DEFINITION / DESCRIPTION**

Acanthamoeba keratitis is a rare eye infection you can get from an amoeba, a microscopic creature similar to bacteria but a little more complex. People who wear contacts or are immunocompromised have the highest risk of contracting this condition. It’s treatable, but the best way to deal with this condition is to prevent it from happening.

Acanthamoeba keratitis (AK) is a rare parasitic infection of the cornea, (the clear dome shaped ‘window’ at the front covering of the eye, that can be very painful) from a certain type of amoeba, when not treated, it can damage the eyes and cause loss of sight. This condition is sometimes known as “amoebic keratitis” or by similar names. AK usually affects one eye at a time, but it can affect both. It starts by affecting the outermost layer of your cornea, the epithelium. As it gets worse, the infection extends deeper.

The infection is caused by a microscopic organism called acanthamoeba, which is common in nature and is usually found in bodies of water (lakes, oceans and rivers) as well as domestic tap water, swimming pools, hot tubs, soil and air.

Many different species of acanthamoeba exist. Acanthamoeba organisms do not generally cause harm to humans (we come into contact with them when we wash, swim, drink water etc), but they can cause a serious eye disease if they infect the cornea. Not all species of acanthamoeba have been found to cause corneal infections. AK is most common in people who wear contact lenses, but anyone with a corneal injury is susceptible to developing the infection.

Acanthamoeba has a life cycle of two stages: an active form (when the organism feeds and replicates), and a dormant form (when the acanthamoeba protects itself from attack by developing into a cyst).

An amoeba is a single-celled organism. It‘s similar to bacteria but a little more complex. It isn‘t a true parasite because it can go through its entire life cycle without needing to infect humans or any other animals. But amoebas act like parasites when they infect humans or animals.

Scientists know of at least 20 species of acanthamoeba worldwide. They can live almost anywhere humans do, and they easily survive in freshwater, seawater, soil and many other places. Researchers know of eight (possibly nine) acanthamoeba species that can cause AK.

During their life cycles, acanthamoeba can take two forms. One is their active, mobile form. The other is their cyst form. This form involves a toughened outer layer. In cyst form, acanthamoeba can survive all kinds of threats that would kill them otherwise.

That includes:

* Extreme temperatures. In cyst form, acanthamoeba can survive temperatures from -4 degrees Fahrenheit to 132 degrees Fahrenheit (-20 degrees Celsius to 56 degrees Celsius).
* Lack of nutrients and water. Acanthamoeba in cyst form can survive up to 20 years (maybe more) at room temperature.
* Chemicals and toxins. Acanthamoeba in cyst form can resist the toxic effects of many substances, including medications that treat parasitic infections.
* Sunlight. The ultraviolet (UV) rays in sunlight can kill many microbes, but acanthamoeba in cyst form can survive.

Knowing that is important because acanthamoeba can enter your body both in active and cyst forms. That’s why you should take proper precautions to prevent them from infecting you.

AK is rare overall. Research indicates there could be up to 1,500 cases in the U.S. each year.

**CAUSES**

**What causes acanthamoeba keratitis (AK)?**

Two species of acanthamoeba cause most cases of AK (experts abbreviate “acanthamoeba” to “A.” in the species names). The two species are *A. castellani* and *A. polyphaga*.

AK is infectious (meaning you can catch it), but it isn’t contagious (you can’t directly catch it from someone else).

The most common ways for acanthamoeba to infect your eyes are:

* Contact lenses.
* Contaminated water.
* Eye injuries.

#### **Contact lenses**

Contact lens wearers make up at least 90% of AK cases. That’s usually because of a combination of factors. The factors include:

* Wearing contacts for too long.
* Improperly storing contact lenses when not wearing them (such as using tap water to clean or store your contacts).
* Incorrectly cleaning contact lenses or the cases you store them in.
* Wearing contact lenses while swimming or showering.
* Using contaminated contact lens-related items, like storage cases or solution.

#### **Contaminated water**

Acanthamoeba can survive in water easily, especially in cyst form. Even treated drinking water, bottled water or swimming pool water may not have a high enough chlorine concentration or other disinfectants. That’s why you should never use tap water with contacts or wear contacts while swimming.

Under ordinary circumstances, acanthamoeba from these sources can’t infect your eyes. But there are times when they can. Some include:

* If you have an existing eye infection.
* If you have an eye injury (like a corneal scratch or similar condition).
* If you’re immunocompromised.

#### **Eye injuries**

The cornea is like your eye‘s windshield, but injuries make it less effective. The injuries are like weak points or small gaps in the cornea surface where it‘s easier for microbes to get in. Injuries to your corneas usually involve contact lenses, your fingers or fingernails, plants or plant material, or dirt or soil.

**RISK FACTORS**

There are a number of different factors which are known to increase the risk of contracting AK. The biggest risk factor is exposure to water (generally through swimming or showering in contact lenses, rinsing or storing lenses in water and handling lenses with unwashed or wet hands). Improper contact lens hygiene, including not disinfecting lenses properly and not cleaning and changing contact lens cases regularly have also been shown to increase the risk of infection. Those who do not wear contact lenses may still contract AK, although it has a much rarer incidence than in contact lens wear.

Due to the way that UK domestic water is stored and supplied, incidence of the disease is generally higher in the UK than in other parts of the world. Studies suggest that AK affects around 2 in 100,000 contact lens wearers per year in the UK, which is around 20 times less than the number of daily wear soft contact lens wearers with bacterial infections. Since 2011, Moorfields Eye Hospital and other centres in the UK and USA have reported a three-fold increase in the number of cases of disease, although it still remains rare.

Risk factors for AK vary by country. In developing countries, trauma remains the leading risk factor for AK and is estimated to be associated with 27% of cases .

**SIGNS / SYMPTOMS**

**What are the symptoms of acanthamoeba keratitis (AK)?**

In the early stages of the disease, the cornea can become irregular due to the infection and inflammation, which can affect your vision.

You may also have light sensitivity (also termed photophobia), which is a symptom of the inflammation and infection in your cornea.

Tears are a natural reaction to disruption of the corneal surface and are a reflex response to the infection.

AK can be extremely painful, although not all patients experience intense pain.

The symptoms of AK happen when the active form of these microbes enters the corneas of your eyes. The symptoms may not remain constant, cycling back and forth between better and worse.

Symptoms include:

* Eye pain (sometimes severe).
* Feeling like something’s stuck in your eye (foreign body sensation), but washing your eyes doesn‘t help, and you can‘t see anything stuck there.
* Watery eye (epiphora).
* Light sensitivity (photophobia).
* Eye redness or irritation.
* Corneas that appear cloudy, dirty or that have a ring-shaped area on their surface.
* Blurred or clouded vision (usually happens with severe or advanced cases).

**DIAGNOSIS METHODS**

AK is tricky to diagnose early on, and an initial misdiagnosis happens in about 75% to 90% of cases. That’s because of the following:

* It has the same symptoms as more common viral or bacterial eye infections.
* AK is rare, so eye care specialists and healthcare providers usually don’t suspect it at first.
* The tests involve a scraping or biopsy of the cornea (which is invasive) or equipment that isn‘t available outside major medical centers.

Your eye care specialist will do an eye exam, including a slit lamp exam. That lets them look into your eyes for signs or clues. They’ll also ask about your symptoms, recent activities or if there could be other contributing factors.

The standard practice is to treat any eye infection as if it were viral or bacterial first and suspect it’s AK if treatment doesn’t work. If it doesn’t, your eye care specialist will likely recommend starting treatment for AK.

**Corneal tissue tests**

Testing for AK may involve taking samples of corneal tissue. That can involve:

* Corneal scraping. To run this test, an eye specialist will take a sample of your cornea’s outermost layers for testing.
* Corneal biopsies. This involves taking a larger tissue sample than with a scraping. The main advantage is that it can detect deeper infections than a scraping test.

Both of these tests are more invasive and can be painful. But your provider will use numbing drops or medications to help with that. Corneas also regenerate quickly, which reduces how long you feel pain or discomfort from a scraping or biopsy.

The wait time for results from corneal tissue testing can take several days. Your provider will likely begin treatment to help with your symptoms in the meantime.

**TREATMENT OPTIONS**

Treating AK has two main goals: getting rid of the infection and reducing pain and other disruptive symptoms. Medications are usually the first option, and surgery may be necessary in more severe cases.

Many of the more common AK medications aren’t available in the United States anymore because the condition is so rare. But some pharmacies may be able to custom-make (compound) these medications for you. Your eye care specialist is the best person to tell you about medication options and where you might be able to get them.

**Medications**

Active forms of acanthamoeba are very sensitive to certain medications. In cyst form, acanthamoeba species can resist treatment, but some medications can still overcome that.

The main form of treatment is topical antiseptic drops, such as chlorhexidine and polyhexanide (also known as polyhexamethylene biguanide, or PHMB).

**Surgery**

About 40% of AK cases don’t respond well enough to medication alone. When that happens, surgery may be the next best option. This can include:

* Epithelial tissue removal (debridement). This involves removing the outermost layer of the cornea, which may remove acanthamoeba living in the cornea (either inactive or cyst form). It can also make it easier for medication to reach acanthamoeba in deeper layers.
* Tissue grafts or special bandages. Certain types of tissue transplants, like amniotic membranes, or a bandage contact lens can help your eye while it heals.
* Keratoplasty and corneal transplants. Removing the cornea may be the only treatment in some cases. Once removed, an eye care specialist can replace it with a cornea transplant.

**PREVENTION TIPS**

AK is mostly preventable, though uncommon cases can happen for reasons you can’t control. Steps you can take include:

* Wear contact lenses as instructed. Don’t wear your contacts for too long, leave them overnight, or wear them while swimming or showering.
* Store your contacts properly. Never use anything but contact lens solution to store your contacts when you take them out. Your eye care specialist can tell you more about the right kind of solution to use.
* Clean your contacts and the case you store them in. Contact lens solution usually doesn’t disinfect your lenses. Your eye care specialist can tell you how to sanitize your contacts and contact lens case, and how often you need to do this.
* Throw contacts away if you get an eye infection and replace the case. Reusing contaminated contacts or contact lens cases can cause reinfection.
* Avoid getting water directly in your eyes. If your eyes feel dry, use artificial tear drops that are intended for use in the eye. If you need to rinse your eyes, contact lens solution is safer than using tap water.
* Take precautions if you’re at higher risk for AK infection. If you have immune system issues or an eye injury that makes you vulnerable to an infection, don’t take chances. Avoid swimming or showering in ways that could allow AK in water (even treated water) to get into your eyes.
* Use eye protection to avoid injuries. AK infections are always a threat with eye injuries. Protecting your eyes keeps them safe from damage, which also keeps microbes out.

**OUTLOOK / PROGNOSIS**

AK is a condition that can be very painful — often much more than your other symptoms or visible eye changes would suggest — and disruptive. It’s important not to ignore it because it gets harder to treat the longer you wait.

If you have AK, your eye care specialist will first try to treat and/or rule out viral and bacterial infections. Make sure you follow up with them and know how long to wait before contacting them again if treatment doesn‘t seem to be helping.

It’s also important to follow your provider’s treatment instructions exactly as guided. That gives you the best chance of a favorable outcome. If you don‘t follow your provider‘s treatment instructions, you may be prone to reinfection. The cyst form can also live dormant in your corneas for extended periods, which can cause AK symptoms to return months after treatment.

AK can cause severe and permanent damage if it goes untreated for too long. The possible complications include glaucoma, iris atrophy (shrinking or moving out of place), cataracts and chronic defects in the cornea’s outermost layer. In more severe cases, major vision loss is possible.

**What’s the outlook for this condition?**

The outlook for AK depends on several factors. Positive outcomes are much more likely if you get medical care before this condition spreads past the second layer of your cornea. That usually means starting treatment within three weeks of when your symptoms start.

You should see an eye care specialist if you have AK symptoms lasting more than a few days. Doing so will allow them time to try bacterial infection treatments, and if those don‘t work, your eye specialist can have you begin AK-specific treatments.

**Living with**

Living with Acanthamoeba Keratitis. Acanthamoeba Keratitis (AK) is a rare condition that occurs when microscopic amoeba invades the cornea – the clear layer - at the front of the eye.

The amoeba normally lives in water and soil and, although the infection has many causes, there is a known risk of infection that can occur when contact lenses come into contact with water because the contact lens can transfer the amoeba to the cornea.

Around 85% of Acanthamoeba Keratitis cases in the UK occur in contact lens wearers.

We want everyone to enjoy the freedom that **contact lenses** offer and give you some basic but vital steps to follow to keep your eyes healthy. Sairia, one of our patients, lost a year of her university course and badly damaged her sight due to poor contact lens care.

The impact of acanthamoeba keratitis on those affected can be extremely difficult. Some patients feel they need additional emotional support, in particular with adjusting to fluctuating vision and changes in appearance as a result of the infection.

**POSSIBLE COMPLICATIONS**

There are a few main complications that are possible with AK:

* Loss of sight. AK damages your corneas, which can lead to loss of sight in the affected eye(s).
* Recurrence. Acanthamoeba in cyst form can live in your corneas for extended periods and return to their live forms later.
* Disruption in daily routine and activities. It‘s usually a painful and disruptive condition. It can interfere with work, hobbies, spending time with loved ones and more.

**WHEN TO SEE A DOCTOR / RED FLAG**

You should see a doctor immediately if you suspect you have Acanthamoeba keratitis, especially if you wear contact lenses and have experienced symptoms such as feeling like something is in your eye, cloudy or dirty-looking corneas, or blurred vision. Prompt medical attention is crucial as the infection can lead to severe pain and potential vision loss if left untreated.

Early diagnosis and treatment are essential for managing the condition effectively.

**DIFFERENTIAL DIAGNOSIS**

Acanthamoeba keratitis can be challenging to diagnose due to its similarity to other types of keratitis. The differential diagnosis includes:

* Herpes Simplex Keratitis: Characterized by dendritic ulcers, this condition can mimic the early stages of Acanthamoeba keratitis. However, Acanthamoeba keratitis typically presents with a more diffuse superficial keratopathy initially, followed by multifocal stromal infiltrates.
* Fungal Keratitis: This condition can resemble Acanthamoeba keratitis in its later stages, particularly with the formation of a ring infiltrate. Fungal keratitis often shows serrated borders, raised slough, and satellite lesions.
* Contact Lens-Associated Keratitis: Wearing contact lenses increases the risk of Acanthamoeba keratitis, making it necessary to consider this condition in patients who wear contact lenses.
* Bacterial Keratitis: While less common, bacterial keratitis can also present with corneal infiltrates and should be considered in the differential diagnosis.
* Sterile Keratitis: Conditions such as those resulting from the use of topical anesthetics can cause keratitis without an identifiable infectious agent, complicating the differential diagnosis.

Each of these conditions requires careful evaluation and appropriate diagnostic testing to distinguish from Acanthamoeba keratitis, including in vivo confocal microscopy, polymerase chain reaction (PCR), and histopathological analysis.

**Can acanthamoeba keratitis be cured?**

Yes, AK is curable. This is easier to do when you get treatment sooner rather than later.

AK is rare overall. You’re more likely to get it if you wear contacts and don’t maintain or store them properly. People with decreased immunity also have a higher risk.

**Is acanthamoeba keratitis an emergency?**

No, AK isn’t an emergency condition. But you need to make an appointment and see an eye care specialist as soon as possible.

There’s no way to tell if you have AK (or any other amoeba-related infection) on your own. Only certain medical tests can show it. If you think you might have AK, you need to see an eye care specialist for diagnosis and treatment.

## Epidemiology of Acanthamoeba Keratitis (AK)

* Global Incidence:  
  The worldwide annual incidence of Acanthamoeba keratitis is estimated at approximately 23,561 cases, which corresponds to about 2.9 cases per million people globally. Another meta-analysis reported a slightly lower estimate of around 2.34 cases per million population, with no significant differences among continents.
* Incidence Among Contact Lens Wearers:  
  Contact lens wear is the major risk factor for AK, especially in high-income countries. The incidence among contact lens users ranges roughly from 1 to 2 new cases per 1 million contact lens wearers annually in the United States. In the Netherlands, the incidence among soft contact lens wearers increased to about 1 in 21,000 by 2015, reflecting a rising trend.
* Geographic Variation:
  + The highest incidence rates are reported in India.
  + Lowest rates have been reported in countries like Tunisia and Belgium.
  + In lower-income regions, ocular trauma, often in agricultural workers, is a more common risk factor than contact lens use.
* Trends:  
  Some countries have reported an increasing incidence over recent years, possibly due to increased contact lens use and improved diagnostic awareness.
* Genotypes:  
  Multiple Acanthamoeba genotypes cause keratitis, with genotype T4 being the most prevalent worldwide.
* Burden and Outcomes:  
  AK is a rare but sight-threatening infection that can lead to permanent blindness if not diagnosed and treated promptly. Treatment failure rates can be as high as 39%, with risk factors including older age, delayed diagnosis, corticosteroid use before diagnosis, and severity at presentation

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**ACHROMATOPSIA**

*Other Names for This Condition* • Achromatism • Rod monochromatism • Total color blindness

**DEFINITION / DESCRIPTION**

Achromatopsia is a condition characterized by a partial or total absence of color vision. People with complete achromatopsia cannot perceive any colors; they see only black, white, and shades of gray. Incomplete achromatopsia is a milder form of the condition that allows some color discrimination. Achromatopsia also involves other problems with vision, including an increased sensitivity to light and glare (photophobia), involuntary back-and-forth eye movements (nystagmus), and significantly reduced sharpness of vision (low visual acuity). Affected individuals can also have farsightedness (hyperopia) or, less commonly, nearsightedness (myopia). These vision problems develop in the first few months of life. Achromatopsia is different from the more common forms of color vision deficiency (also called color blindness), in which people can perceive color but have difficulty distinguishing between certain colors, such as red and green. Frequency Achromatopsia affects an estimated 1 in 30,000 people worldwide. Complete achromatopsia is more common than incomplete achromatopsia. Complete achromatopsia occurs frequently among Pingelapese islanders, who live on one of the Eastern Caroline Islands of Micronesia. Between 4 and 10 percent of people in this population have a total absence of color vision

**Achromatopsia**

Achromatopsia is an inherited vision disorder that limits your ability to see color. It’s present at birth and usually nonprogressive, meaning the symptoms don't worsen over time.

***Types of achromatopsia***

There are two types:

* Complete: Vision is limited to black, white and shades of grey.
* Incomplete: Color vision is limited, with dull hues that can be difficult to distinguish.

**CAUSES OF ACHROMATOPSIA**

Achromatopsia results from changes in one of several genes: CNGA3, CNGB3, GNAT2, PDE6C, or PDE6H. A particular CNGB3 gene mutation underlies the condition in Pingelapese islanders. Achromatopsia is a disorder of the retina, which is the light-sensitive tissue at the back of the eye. The retina contains two types of light receptor cells, called rods and cones. These cells transmit visual signals from the eye to the brain through a process called phototransduction. Rods provide vision in low light (night vision). Cones provide vision in bright light (daylight vision), including color vision. 1 Reprinted from MedlinePlus Genetics (https://medlineplus.gov/genetics/) Mutations in any of the genes listed above prevent cones from reacting appropriately to light, which interferes with phototransduction. In people with complete achromatopsia, cones are nonfunctional, and vision depends entirely on the activity of rods. The loss of cone function leads to a total lack of color vision and causes the other vision problems. People with incomplete achromatopsia retain some cone function. These individuals have limited color vision, and their other vision problems tend to be less severe. Some people with achromatopsia do not have identified mutations in any of the known genes. In these individuals, the cause of the disorder is unknown. Other genetic factors that have not been identified likely contribute to this condition.

**RISK FACTORS OF ACHROMATOPSIA**

Achromatopsia is primarily a genetic condition, typically inherited in an autosomal recessive manner, meaning both parents must carry the gene for their child to be affected.

Risk factors include having parents who are carriers of the gene responsible for achromatopsia. If both parents are carriers, each child has a 25% chance of being affected, a 50% chance of being a carrier, and a 25% chance of neither being affected nor a carrier.

The condition is more prevalent in populations where marriages among close relatives are common, as this increases the likelihood of both partners carrying the same recessive gene.

An example of this is seen on the island of Pingelap in Micronesia, where a typhoon in the late 1700s led to a founder effect, causing a high prevalence of achromatopsia, with up to 10% of the population affected.

Genetic testing can help identify carriers and assess the risk for future generations.

**Risk to Family Members**

*Parents of a proband*

* The parents of an affected child are obligate heterozygotes (i.e., carriers of one achromatopsia-related pathogenic variant).
* Heterozygotes (carriers) are asymptomatic and are not at risk of developing the disorder.

*Sibs of a proband*

* At conception, each sib of an affected individual has a 25% chance of being affected, a 50% chance of being an asymptomatic carrier, and a 25% chance of being unaffected and not a carrier.
* Heterozygotes (carriers) are asymptomatic and are not at risk of developing the disorder.

Offspring of a proband. The offspring of an individual with achromatopsia are obligate heterozygotes (carriers) for an achromatopsia-related pathogenic variant.

Other family members. Each sibling of the proband's parents is at a 50% risk of being a carrier of an achromatopsia-related pathogenic variant.

**SIGNS / SYMPTOMS**

Achromatopsia is characterized by several symptoms, including extreme sensitivity to bright light, also known as hemeralopia, poor visual acuity, and a lack of color vision.

Affected individuals often experience nystagmus, which is an involuntary, repetitive movement of the eyes.

These symptoms typically become noticeable in early childhood, often around three to six months of age.

Additionally, patients with achromatopsia frequently require glasses to correct hyperopia, or farsightedness.

**Symptoms of achromatopsia**

With achromatopsia, you may experience:

* Blind spots (scotomas).
* Blurred vision (astigmatism).
* Color blindness.
* Extreme farsightedness.
* Eye discomfort in bright light (photophobia).
* Myopia (nearsightedness).
* Poor or low vision.
* Rapid eye movements (nystagmus).

**How soon do children experience symptoms?**

Light sensitivity occurs in the first months of life. Symptoms, such as poor vision and color blindness, may also be present. But you might not notice them until your child is a little older.

**DIAGNOSIS METHODS**

Achromatopsia should be suspected in individuals with the following typical clinical findings, additional testing, and family history.

An eye care professional (ophthalmologist) diagnoses achromatopsia. The assessment starts by reviewing your family history and symptoms.

A retinal exam may be normal, so additional tests are necessary. These include:

* Color vision testing, which assesses your ability to distinguish different colors.
* Fundus autofluorescence, which uses blue light to examine tissue in the back of the eye (retina).
* Ophthalmic electrophysiology, which evaluates how your eyes and supporting nerves respond to light.
* Electroretinography (ERG), a component of ophthalmologic electrophysiology, this test measures the electrical response of rods and cones.
* Optical coherence tomography (OCT), which generates images of the retina.
* Visual field testing, which shows whether you have blind spots and, if so, how large they are.

**TREATMENT OPTIONS**

**Management**

Achromatopsia has no cure. People are still able to lead an independent life by maximizing available vision, social support, and managing symptoms.

#### **Special glasses**

Treatment often includes dark-tinted glasses. The lenses filter out specific types of light. Frames may extend toward the temples to maximize coverage. They may also have a shield at the top.

#### **Low vision therapy**

You learn how to complete daily tasks safely by:

* Making materials easier to read with help from electronic magnification devices.
* Navigating unfamiliar places with the help of a long white cane.
* Scanning surroundings for potential fall hazards.
* Taking public transportation to get around if you cannot drive.
* Using high-contrast materials, such as black ink on white paper, that make things easier to see.

**PREVENTION TIPS**

There isn’t anything you can do to prevent this condition. If it runs on both sides of your family, you may wish to consider genetic testing. The results let you know the chances of passing on the condition to your children.

## **Living With**

Certain methods and habits can help you stay safe, comfortable and maximize independence. These include:

**Home organization and decor**

* Arrange the furniture in your home to maximize open space and limit the likelihood of bumping into things.
* Hang thick curtains in your home that let you control the amount of natural light.
* Minimize glare on surfaces like your walls by using a matte paint.
* Organize your home in ways that make it easier to find essential items. This may include labeling items with tags that have a large, thick font.

**Daily activities**

* Avoid bright light “whiteouts” by not leaving the house in the middle of the day.
* Get a screen reader to avoid looking at electronic device displays that may be too bright.
* Use vision aid technologies, such as a handheld scanner that announces the object’s color.
* Wear a brimmed hat while outside.

# **OUTLOOK / PROGNOSIS**

The prognosis is good:

* Children typically attend regular school. Achromatopsia does not cause learning issues. But children may need assistance to overcome vision-related challenges.
* Adults with achromatopsia often live independently. They may need ongoing support to adapt to their environment and daily activities.

**POSSIBLE COMPLICATIONS**

Achromatopsia is a rare inherited condition that primarily affects the cone photoreceptors, leading to several complications. These include:

* Photophobia: Extreme sensitivity to bright light, making it difficult for individuals to tolerate sunlight or bright indoor lighting.
* Nystagmus: Involuntary rhythmic eye movements that can occur in response to light sensitivity or as a result of the condition itself.
* Reduced Visual Acuity: Blurry vision, typically around 20/200, which does not significantly improve with age.
* Day Blindness: Difficulty seeing clearly in bright light conditions, while vision may be better adapted to dimmer environments.

These symptoms can significantly impact daily activities and quality of life. However, the condition is considered stable, with progression over time being relatively slow and subtle.

**WHEN TO SEE A DOCTOR / RED FLAG**

Individuals with suspected achromatopsia should consult an eye doctor for a thorough evaluation and diagnosis. Symptoms of achromatopsia include difficulty seeing colors, reduced visual acuity, extreme light sensitivity, and nystagmus (involuntary eye movements).

An eye doctor can perform several tests to confirm the presence of achromatopsia, including checking for color vision, visual acuity, and examining the retina.

Additionally, genetic testing may be recommended to identify the specific genetic mutation causing the condition.

Early diagnosis and intervention can help manage symptoms and improve quality of life. Treatment options may include the use of tinted lenses, low vision aids, and adaptive strategies to cope with light sensitivity and other visual impairments.

* Tinted Lenses: Special glasses with tinted lenses can help filter out specific types of light, reducing glare and improving vision in bright conditions.
* Low Vision Aids: Devices such as digital magnifiers can assist with tasks like reading and using computers.
* Adaptive Strategies: Learning techniques to navigate and adapt to environments with varying light conditions can also be beneficial.

Consultation with a low vision optometrist can provide personalized recommendations based on individual needs and lifestyles.

**DIFFERENTIAL DIAGNOSIS**

Other conditions that may present with similar symptoms include:

* Cone Dystrophy: A group of inherited retinal disorders affecting cone cells.
* Rod-Cone Dystrophy: A condition that affects both rod and cone photoreceptors.
* Retinitis Pigmentosa: A genetic disorder that leads to progressive vision loss.

**How is achromatopsia different from color blindness?**

In color blindness, people have normal vision and see some color. In achromatopsia, vision is reduced, there is a lack of color vision, and other vision issues arise such as rapid eye movements. Symptoms often make it difficult to go about daily life.

**RECENT GUIDELINES, UPDATES, AND RESEARCH**

## Current Management Recommendations

* Symptomatic Management
  + Use of tinted or sunglasses to reduce photophobia and improve comfort in bright environments.
  + Low vision aids such as magnifiers and specialized glasses to enhance residual vision.
  + Correction of refractive errors with appropriate lenses.
  + Environmental and lifestyle modifications to reduce glare and optimize lighting.
  + Nutritional support with a balanced diet rich in antioxidants and nutrients to support overall eye health.
* Regular Monitoring
  + Routine eye examinations including visual acuity, color vision testing, and retinal imaging (OCT) to monitor disease progression.
  + Electroretinography (ERG) remains the gold standard diagnostic tool to assess cone function.

## Advances in Gene Therapy

* Gene Replacement Therapy targeting mutations in CNGA3 and CNGB3 genes, the most common causes of achromatopsia, has shown encouraging results in early-phase clinical trials.
* Trials at institutions such as University College London and Casey Eye Institute have demonstrated partial restoration of cone function and improvements in color vision in some patients, including children.
* Ongoing Phase I/II studies are assessing safety and efficacy of subretinal injections of AAV vectors expressing functional CNGA3 or CNGB3 genes.

## Clinical Trial Eligibility Criteria (Example from CNGB3 Gene Therapy Trial)

* Clinical diagnosis of achromatopsia with documented CNGB3 mutation.
* Visual acuity no better than 20/80 in the treated eye.
* Age criteria vary; some trials include children as young as 6 years.
* Good general health without other ocular diseases.

## Research and Diagnostic Innovations

* Comprehensive phenotyping using OCT and ERG helps stratify patients and monitor structural and functional changes.
* Identification of novel genetic variants expands understanding of disease heterogeneity and informs personalized approaches.

**EPIDEMIOLOGY**

Achromatopsia affects an estimated 1 in 30,000 people worldwide. Complete achromatopsia is more common than incomplete achromatopsia.

Complete achromatopsia occurs frequently among Pingelapese islanders, who live on one of the Eastern Caroline Islands of Micronesia. Between 4 and 10 percent of people in this population have a total absence of color vision.

Achromatopsia affects approximately 1 in 30,000 people around the world. It is more common in areas where marriages among close relatives are more likely to occur.

It is also more prevalent in Pingelap, a small group of islands in the eastern Pacific Ocean. As described in the Oliver Sacks book, “ The Island of the Colorblind,” a typhoon in the 1700s affected the population of the islands. This left a small number of people to repopulate the area and caused the propagation of an achromatopsia-carrying gene.

**What are my chances of having achromatopsia?**

You are more likely to have this condition if there’s a family history. If achromatopsia runs on both sides of your family, your chances of having it are 1 in 4.

*REFERENCE:*

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**ADIE’S PUPIL**

*Adie’s pupil is also known as tonic pupil or Holmes-Adie pupil.*

**DEFINITION / DESCRIPTION**

This is a neurological disorder—a type of disease that affects the nervous system. The nervous system—made up of the brain, spinal cord, and nerves—controls many of our involuntary bodily functions. These are reflexive actions that happen automatically, without having to think about them—things like sweating, salivating, and sneezing.

The nervous system also controls the pupil (small hole in the center of the iris) and its response to light. Normally, the pupil constricts (gets smaller) in brighter light to let less light in. In lower light, the pupil dilates (widens) to let more light in, so we can see better.

With Adie’s pupil, there is an abnormal pupillary response to light. In most cases, it affects only one eye. The affected pupil is usually larger than normal and does not constrict as it should in the presence of bright light.

**CAUSES**

No one knows for sure what causes Adie’s pupil. Most doctors think it’s caused by a viral or bacterial infection that damages the nerves that control the pupil. Some think it may be caused by autoimmune disease, when the body’s immune system attacks its own healthy tissues, like the nerves that operate the pupil.

**RISK FACTORS**

Adie's pupil, also known as Adie's tonic pupil, is a condition characterized by a dilated pupil that reacts slowly to light and accommodates poorly. The exact cause of Adie's pupil is often unknown, but several risk factors and associated conditions have been identified:

* Viral infections, particularly neurosyphilis, have been linked to Adie's pupil.
* Trauma or surgery involving the eye area can lead to the development of Adie's pupil.
* Lack of blood flow (ischemia) to the eye can contribute to the condition.
* Autoimmune processes may also play a role in the development of Adie's pupil.

These factors can cause inflammation or damage to the ciliary ganglion, a cluster of nerve cells in the eye socket that controls the pupil's response to light and accommodation.

**SYMPTOMS / SIGNS**

Symptoms of Adie’s pupil can include having:

* a pupil that is larger than the other
* a pupil that doesn’t get smaller in bright light
* light sensitivity
* blurry vision
* difficulty reading (with Adie’s pupil, the eye has a hard time focusing for near tasks)

Rarely, both eyes are affected. And sometimes, Adie’s pupil has the opposite effect on a patient’s pupil(s), where they fail to widen adequately in low light situations.

There are also some non-eye related symptoms that are common with Adie’s pupil, including:

* excessive sweating
* not having a knee-jerk reflex

## **Additional symptoms**

In addition to ocular symptoms, Adie syndrome is characterized by sluggish or missing deep tendon reflexes. Deep tendon reflexes are muscle reactions that are automatic and involuntary, such as knee reflexes and the Achilles tendon reflex.

Additional symptoms that a person may experience include:

* Headaches
* Pain in the face
* Mood swings

A person may experience changes in sweating, which can be excessive or reduced in some circumstances.

**Ross’s Syndrome:** While Ross’s syndrome is technically the combination of decreased sweating, missing reflexes, and weak pupil responses, some clinicians may describe this condition as a variant of Adie syndrome.

Although the symptoms of Adie syndrome are rarely severe or disabling, they can cause significant disruption in everyday life and require treatment.

**DIAGNOSIS METHOD**

Adie’s pupil can usually be diagnosed during an eye exam with your ophthalmologist. The exam may include:

* **Special diagnostic eye drops**. Your ophthalmologist gives you these drops to see how the [pupil](https://www.aao.org/eye-health/anatomy/pupil) responds. A pupil with Adie’s will get smaller after using these drops.
* **A slit-lamp exam**. This device magnifies and illuminates your pupils. Seen close-up, the pupils may show signs of Adie’s.
* **Pupil response testing**. Your doctor will want to see how your pupil responds to bright light and low light. These responses are then compared to the unaffected eye. They may also test to see how the pupil accommodates, or focuses, on an object placed very close to the eye.

In some cases, your ophthalmologist will want you to see a neurologist. A neurologist is a doctor with special training in diseases of the nervous system.

**TREATMENT OPTIONS**

There is no cure for Adie’s pupil, but there are ways to relieve some of the symptoms. Your doctor may suggest:

* glasses to improve reading or near vision
* sunglasses to reduce light sensitivity
* eye drops to make pupil(s) smaller and reduce light sensitivity. Eye drops can also reduce glare while driving at night.

Depending on the cause, some people with Adie’s may recover their normal pupillary response. In others, pupillary function is never recovered or never fully recovered. It is helpful to know the disease is not life-threatening, and with proper treatment, those with Adie’s pupil can manage their condition and expect to live full and healthy lives.

**PROGNOSIS**

Adie syndrome does not have a progressive course. It is not a life-threatening condition and does not cause disabilities. It is not associated with any mortality rate. The loss of deep tendon reflexes is permanent and may progress over time. Most patients require reassurance after confirmation of the diagnosis. There have been rare associations of angle-closure glaucoma with Adie pupil. The accommodative paresis gets better spontaneously over time. The pupil light reaction becomes weaker over time with an increasing light-near dissociation, and the pupil becomes smaller with time ("little old Adie")

**POSSIBLE COMPLICATION**

Adie syndrome may rarely result in angle-closure glaucoma that leads to episodes of blurring of vision and ocular pain. Management includes medical treatment to lower the intraocular pressure and laser iridotomy to prevent pupillary block.

Previously, Adie syndrome has also been reported to result in amblyopia in children due to the conversion of latent hypermetropia into manifest hypermetropia as a result of accommodative paresis. This has been effectively managed by correction of the refractive error and occlusion therapy.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnoses of Adie syndrome include other causes of light-near dissociation such as Argyll Robertson pupil that is found in late-stage syphilis and is associated with miosis. Other causes of mydriasis including pharmacological dilation which lacks light-near dissociation, oculomotor nerve palsy which may be associated with ophthalmoplegia and ptosis, and optic nerve diseases which are associated with a relative afferent pupillary defect and diminution of vision should be excluded.

Systemic autonomic neuropathies like Ross and harlequin syndromes can also affect the ciliary ganglion and produce the tonic pupil. Ross syndrome is characterized by a triad of a tonic pupil, hyporeflexia, and segmental anhidrosis. Harlequin syndrome is a rare disorder of the sympathetic nervous system characterized by the unilateral decrease or absence of flushing and sweating with an exaggerated response on the contralateral normal side, particularly in the face, neck, arm, and chest in response to heat, exercise, or emotional factors.Some patients (13% in one series) also have a tonic pupil, although a Horner syndrome is more common and may coexist with a tonic pupil.

Little old Adie with a miotic pupil should be differentiated from other causes of miosis. Horner syndrome is another cause of a miotic pupil, however, ptosis and apparent enophthalmos are usually present. Miosis may also be present as an early sign in temporal lobe herniation following head injury due to oculomotor nerve irritation, which is known as Hutchison's stage I. This, however, is usually followed by pupillary dilatation.

Adie’s pupil needs to be distinguished from pupil abnormalities caused by the following factors:

**Oculomotor nerve palsy**

In addition to the dilated pupils and loss of light reflection, there are other clinical manifestations, such as ptosis and restricted eye movements. Brain magnetic resonance imaging can reveal damage to the oculomotor nerve nucleus and fibers , and the pilocarpine test is usually negative.

**Drug overdose**

Atropine is an anticholinergic drug that can lead to a series of anticholinergic symptoms after excessive intake, including dilated pupils, hallucinations, agitation, tachycardia, delirium, and fever. In addition, Datura, a traditional Chinese medicine, can also cause the above symptoms when ingested in large quantities.

**Argyll-Robertson pupil**

It is generally thought to be related to neurosyphilis. Patients usually present with bilateral miosis, loss of direct and indirect light reflection, and presence of accommodation and convergence reflexes. The affected pupil dilated after atropine instillation.

**Congenital mydriasis**

Mydriasis and associated loss of light reflex are congenital, and both pupils can be affected. It is more common in women, and the pathogenesis is unclear. A combination of medical history and negative pilocarpine test can assist in identification of congenital mydriasis.

**RECENT GUIDELINES OR UPDATES**

## Management and Treatment Recommendations

* General Approach:
  + Adie pupil is benign and often requires no treatment.
  + Patient reassurance regarding the non-life-threatening nature of the condition.
* Symptomatic Treatment:
  + Sunglasses to reduce photophobia and light sensitivity.
  + Reading glasses or bifocals for accommodative difficulties.
  + Dilute pilocarpine eye drops (0.125%) can be used to constrict the pupil, reduce photophobia, and improve near vision. However, side effects include brow ache, ciliary spasm, and potential worsening of anisocoria.
  + Use of eye drops is generally reserved for patients with significant symptoms.
* Referral:
  + Ophthalmology for diagnosis and management.
  + Neurology referral if systemic neurological symptoms or atypical presentation occur.
* Prognosis:
  + Accommodative paresis may improve over months to years.
  + Pupillary light response usually does not fully recover.
  + Condition is chronic but non-progressive and not life-threatening.

**EPIDEMIOLOGY**

The incidence of Adie syndrome is approximately 4.7/100,000 population/year with a prevalence of two cases/1000 population (approximately). Young adults usually between the ages of 25 to 45 (mean age of 32 years) are most commonly affected. There is a female predominance (2.6:1). Sporadic Adie syndrome is commonly reported with rare reports of familial association. It is unilateral in 80% of the cases. The exact incidence and prevalence rates have not been reported.

Adie syndrome is a relatively rare condition affecting 5 in every 100,000 people, most often young adults aged 25-45.

Adie syndrome has multiple names:

* Adie’s Pupil
* Adie’s Syndrome
* Adie’s Tonic Pupil
* Tonic pupil syndrome
* Holmes-Adie Syndrome (HAS)

*REFERENCES:*

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**AGE-RELATED MACULAR DEGENERATION (AMD)**

Age-related macular degeneration is also known as macular degeneration. **DEFINITION / DESCRIPTION**

This is a disease that damages the sharp and central vision, affecting the ability to see objects clearly, read, or drive. Degeneration of the central retina (macula) causing loss of central vision, common in older adults.

**Two Types of Macular Degeneration**

There are two types of Macular degeneration:

1. **WET (EXUDATIVE) AGE-RELATED MACULAR DEGENERATION**

Wet macular degeneration is an eye condition that causes blurred vision or reduced central vision. It is a type of age-related macular degeneration where blood vessels leak fluid or blood into part of the retina known as the macula (MAK-u-luh). The macula is responsible for central vision.

Wet macular degeneration is one of two types of age-related macular degeneration. The other type, dry macular degeneration, is more common and less severe. The wet type always begins as the dry type.

Early detection and treatment of wet macular degeneration may help reduce vision loss. In some instances, early treatment may recover vision.

This form is less common but much more serious. Wet AMD is when new, abnormal blood vessels grow under the retina. These vessels may leak blood or other fluids, causing scarring of the macula. You lose vision faster with wet AMD than with dry AMD.

Many people don’t realize they have AMD until their vision is very blurry. This is why it is important to have regular visits to an ophthalmologist. They can look for early signs of AMD before you have any vision problems.

**CAUSES**

Wet age-related macular degeneration (AMD) occurs when abnormal blood vessels grow into the macula, a small but vital part of the retina responsible for central vision.

These new blood vessels, known as choroidal neovascularization (CNV), can leak fluid or blood, leading to scarring of the macula and rapid loss of central vision.

The exact cause of wet AMD is not fully understood, but both genetic and environmental factors are thought to play a role.

Research is ongoing to understand the underlying mechanisms and to develop new treatments for wet AMD.

For instance, scientists are exploring the use of stem cells to treat dry AMD in animals, which could potentially lead to treatments for wet AMD in the future.

No one knows the exact cause of wet macular degeneration, but it develops in people who have dry macular degeneration. Of all people with age-related macular degeneration, about 20% have the wet form.

Wet macular degeneration can develop in different ways:

* **Vision loss caused by irregular blood vessel growth.** Sometimes new blood vessels grow from the choroid under and into the macula. This growth isn't typical, and when it happens it's known as choroidal neovascularization. The choroid is the layer of blood vessels between the retina and the outer, firm coat of the eye, called the sclera. These blood vessels may leak fluid or blood, affecting the retina's function and leading to vision loss.
* **Vision loss caused by fluid buildup in the back of the eye.** When fluid leaks from the choroid, it can collect between the thin cell layer called the retinal pigment epithelium and the retina or within the layers of the retina. This may cause irregularities in the macula layers, resulting in vision loss or distortion.

**RISK FACTORS**

Factors that may increase the risk of macular degeneration include:

* **Age.** This disease is most common in people over 50.
* **Family history and genetics.** This disease has a hereditary component, meaning it can run in families. Researchers have identified several genes linked to the condition.
* **Race.** Macular degeneration is more common in white people.
* **Smoking.** Smoking cigarettes or being exposed to tobacco smoke on a regular basis greatly increases the risk of macular degeneration.
* **Obesity.** Research suggests that obesity may increase the chance that early or intermediate macular degeneration will progress to a more severe form of the disease.
* **Cardiovascular disease.** If you have diseases that affect your heart and blood vessels, you may be at higher risk of macular degeneration.

**SIGNS / SYMPTOMS**

Wet macular degeneration symptoms usually appear suddenly and worsen quickly. They may include:

* Visual distortions, such as straight lines that seem to be bent.
* Reduced central vision in one or both eyes.
* The need for brighter light when reading or doing close-up work.
* Difficulty adjusting to low light levels, such as when entering a dimly lit restaurant or theater.
* Increased blurriness of printed words.
* Difficulty recognizing faces.
* A well-defined blurry spot or blind spot in the field of vision.

Macular degeneration doesn't affect side vision, so it doesn't cause total blindness.

**DIAGNOSIS METHODS**

To diagnose wet macular degeneration, an eye doctor typically reviews medical and family history and does a complete eye exam. To confirm a diagnosis of macular degeneration, an eye doctor may suggest other tests, including:

* **Examination of the back of the eye.** An eye doctor puts drops in the eyes to dilate them and uses a special tool to examine the back of the eye. The eye doctor looks for a mottled appearance that's caused by yellow deposits that form under the retina, called drusen. People with macular degeneration often have many drusen.
* **A test for changes in the center of the vision field.** An Amsler grid may be used to test for changes in the center of the vision field. In macular degeneration, some of the straight lines in the grid may look faded, broken or distorted.
* **Fluorescein angiography.** During this test, an eye doctor injects a dye into a vein in the arm. The dye travels to and highlights the blood vessels in the eye. A special camera takes pictures as the dye travels through the blood vessels. The images may show leaking blood vessels or retinal changes.
* **Indocyanine green angiography.** Like fluorescein angiography, this test uses an injected dye. It may be used to confirm the findings of a fluorescein angiography or to identify problem blood vessels deeper in the retina.
* **Optical coherence tomography.** This noninvasive imaging test displays detailed cross sections of the retina. It identifies areas of thinning, thickening or swelling. This test also is used to help monitor how the retina responds to macular degeneration treatments.
* **Optical coherence tomography (OCT) angiography.** This noninvasive imaging test displays detailed cross sections of the retina. It identifies areas of thinning, thickening or swelling. These can be caused by fluid buildup from leaking blood vessels in and under the retina.

**TREATMENT OPTIONS**

Treatments are available that may help slow disease progression and preserve existing vision. If started early enough, treatment may recover some lost vision.

**Medicines**

Some medicines, called anti-VEGF drugs, may help stop the growth of new blood vessels. These medicines block the effects of growth signals the body sends to generate new blood vessels. They are considered the first line of treatment for all stages of wet macular degeneration.

Medicines used to treat wet macular degeneration include:

* Bevacizumab (Avastin).
* Ranibizumab (Lucentis).
* Aflibercept (Eylea).
* Brolucizumab (Beovu).
* Faricimab-svoa (Vabysmo).

An eye doctor injects these medicines into the affected eye. Shots may be needed every 4 to 6 weeks to maintain the beneficial effect of the medicine. In some instances, vision may be partially recovered as the blood vessels shrink and the body absorbs the fluid under the retina.

Possible risks of these shots include:

* Conjunctival hemorrhage.
* Increased eye pressure.
* Infection.
* Retinal detachment.
* Eye inflammation.

**Therapies**

* **Photodynamic therapy.** This procedure is a possible treatment for the irregular blood vessel growth in wet macular degeneration. However, it is much less common than treatment with anti-VEGF shots.  
  During photodynamic therapy, an eye doctor injects a medicine called verteporfin (Visudyne) into a vein in the arm. The medicine then travels to blood vessels in the eye. An eye doctor shines a focused light from a special laser on the affected blood vessels in the eye. This activates the verteporfin, causing the blood vessels to close. This stops the leakage.  
  Photodynamic therapy may improve vision and reduce the rate of vision loss. Repeated treatments may be needed over time, as the treated blood vessels may reopen.  
  After photodynamic therapy, it may be necessary to avoid direct sunlight and bright lights until the medicine has cleared the body. This may take a few days.
* **Photocoagulation.** During photocoagulation therapy, an eye doctor uses a high-energy laser beam to seal problem blood vessels under the macula. This procedure helps stop the vessels from bleeding, with the aim of minimizing further damage to the macula. Even with this treatment, blood vessels may regrow, requiring further treatment. The laser also can cause scarring that creates a blind spot.  
  Few people who have wet macular degeneration get this treatment. It generally isn't an option if you have problem blood vessels directly under the center of the macula. Also, the more damaged the macula is, the lower the likelihood of success.
* **Low vision rehabilitation.** Age-related macular degeneration doesn't affect side vision and typically doesn't cause total blindness. But it can reduce or eliminate central vision. You need central vision to read, drive and recognize people's faces. It may help to get care from a low vision rehabilitation specialist, an occupational therapist, an eye doctor and others trained in low vision rehabilitation. They can help find ways to adapt to changing vision.

**PREVENTION TIPS**

It's important to have routine eye exams to identify early signs of macular degeneration. The following measures may help reduce the risk of developing wet macular degeneration:

* **Manage all other medical conditions.** For example, if you have cardiovascular disease or high blood pressure, take your medicine and follow your healthcare team's instructions for controlling the condition.
* **Don't smoke.** People who smoke are more likely to develop macular degeneration than are people who don't smoke. Ask a healthcare professional for help stopping smoking.
* **Maintain a healthy weight and exercise regularly.** If you need to lose weight, reduce the number of calories you eat and increase the amount of exercise you get each day.
* **Choose a diet rich in fruits and vegetables.** These foods contain antioxidant vitamins that reduce your risk of developing macular degeneration.
* **Include fish in your diet.** Omega-3 fatty acids, which are found in fish, may reduce the risk of macular degeneration. Nuts such as walnuts also contain omega-3 fatty acids.

**Lifestyle and home remedies**

Even after you get a diagnosis of wet macular degeneration, you can take some steps that may help slow vision loss.

* **Don't smoke.** If you smoke, ask a healthcare professional for help quitting.
* **Choose a healthy diet.** The antioxidant vitamins in fruits and vegetables contribute to eye health. Kale, spinach, broccoli, squash and other vegetables have high levels of antioxidants, including lutein and zeaxanthin. These nutrients may benefit people with macular degeneration.  
  Eating foods with high levels of zinc also may be helpful for people with macular degeneration. These include high-protein foods, such as beef, pork and lamb. Non Meat sources include milk, cheese, yogurt, whole-grain cereals and whole-wheat bread.  
  Another good choice is healthy unsaturated fat, such as in olive oil. And research studies have shown that a diet high in omega-3 fatty acids, such as in salmon, tuna and walnuts, may lower the risk of advanced macular degeneration. But the same benefit is not shown from taking omega-3 supplements, such as fish oil pills.
* **Manage your other medical conditions.** If you have cardiovascular disease or high blood pressure, for example, take your medicine and follow your healthcare team's instructions for controlling the condition.
* **Maintain a healthy weight and exercise regularly.** If you need to lose weight, reduce the number of calories you eat and increase the amount of exercise you get each day.
* **Have routine eye exams.** Ask your eye doctor about the recommended schedule for follow-up exams. In between checkups, you can do a self-assessment of your vision using an Amsler grid.

**Vitamin supplements**

For people with intermediate or advanced disease, taking a high-dose formulation of antioxidant vitamins and minerals may help reduce the risk of vision loss. Research from the Age-Related Eye Disease Study 2 (AREDS2) has shown benefit in a formulation that includes:

* 500 milligrams (mg) of vitamin C.
* 400 international units (IU) of vitamin E.
* 10 mg of lutein.
* 2 mg of zeaxanthin.
* 80 mg of zinc as zinc oxide.
* 2 mg of copper as cupric oxide.

Ask your eye doctor if taking supplements is right for you.

Vision loss from macular degeneration can affect the ability to do things such as read, recognize faces and drive. These tips may help to cope with changing vision:

* **Get your eyeglass prescription checked.** If you wear contacts or glasses, be sure your prescription is up to date. If new glasses don't help, ask for a referral to a low vision specialist.
* **Use magnifiers.** A variety of magnifying devices can help you with reading and other close-up work, such as sewing. Such devices include hand-held magnifying lenses or magnifying lenses you wear like glasses.  
  You also may use a closed-circuit television system that uses a video camera to magnify reading material and project it on a video screen.
* **Change your computer display and add audio systems.** Adjust the font size in your computer's settings. And adjust your monitor to show more contrast. You also may add speech-output systems or other technologies to your computer.
* **Use electronic reading aids and voice interfaces.** Try large-print books, tablet computers and audiobooks. Some tablet and smartphone apps are designed to help people with low vision. And many of these devices now come with voice recognition features.
* **Select special appliances made for low vision.** Some clocks, radios, telephones and other appliances have extra-large numbers. You may find it easier to watch a television with a larger high-definition screen, or you may want to sit closer to the screen.
* **Use brighter lights in your home.** Better lighting helps with reading and other daily activities, and it may reduce the risk of falling.
* **Consider your transportation options.** If you drive, check with your doctor to see if it's safe to continue doing so. Be extra cautious in certain situations, such as driving at night, in heavy traffic or in bad weather. Use public transportation or ask a friend or family member to help, especially with night driving. Or use local van or shuttle services, volunteer driving networks, or ride-sharing.
* **Get support.** Having macular degeneration can be difficult, and you may need to make changes in your life. You may go through many emotions as you adjust. Consider talking to a counselor or joining a support group. Spend time with supportive family members and friends.

**POSSIBLE COMPLICATIONS**

People whose wet macular degeneration has progressed to central vision loss have a higher risk of depression and social isolation. With profound loss of vision, people may see visual hallucinations. This condition is known as Charles Bonnet syndrome.

**WHEN TO SEE A DOCTOR / RED FLAG**

See your eye care professional if:

* You notice changes in your central vision.
* You lose the ability to see fine detail.

These changes may be the first sign of macular degeneration, particularly if you're older than age 60.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis of wet age-related macular degeneration (AMD) includes conditions that can also cause choroidal neovascularization (CNV). These conditions include ocular histoplasmosis syndrome, pathologic myopia, choroidal rupture, angioid streaks, and idiopathic causes. Additionally, foveal detachments or vitelliform detachments may complicate other conditions such as cuticular drusen or pattern dystrophies.

Other potential causes of CNV that need to be considered in the differential diagnosis include conditions such as central serous chorioretinopathy (CSC), pattern dystrophy, and drug toxicity, although these are more commonly associated with non-exudative (dry) AMD.

Breakthrough vitreous hemorrhage can also occur in wet AMD, and diagnosis may be challenging if the view is obscured during a dilated retinal examination. In such cases, evaluating the fellow eye or obtaining a detailed history can help establish the diagnosis.

Regular monitoring is crucial for assessing treatment effectiveness and making the necessary adjustments

**RECENT GUIDELINES OR UPDATES**

**Wet AMD Guidelines**

Recent guidelines for the management of wet age-related macular degeneration (wAMD) emphasize the importance of early detection and prompt treatment to prevent severe vision loss. According to the NCBI Bookshelf guidelines published in January 2018, early AMD is defined as having no signs of AMD or small ('hard') drusen (less than 63 micrometres).

For late AMD (dry), fundus examination is recommended for confirmation.

For wet AMD, the guidelines recommend offering optical coherence tomography (OCT) to people with suspected late AMD (wet active) and making an urgent referral to a macula service within one working day if neovascularization is suspected.

Anti-vascular endothelial growth factor (anti-VEGF) treatments are recommended for late AMD (wet active) and should be administered as soon as possible, ideally within 14 days of referral.

The guidelines also highlight the importance of monitoring for depression in patients with AMD due to the increased risk associated with the condition.

Additionally, the guidelines suggest considering referral to low-vision services and group-based rehabilitation programs to promote independent living for people with AMD.

A panel of Greek experts proposed guidelines for the management of wAMD, focusing on defining successful treatment and non-response to therapy. Successful responses to treatments include any gain in best-corrected visual acuity (BCVA) or vision loss that is less than 5-10 Early Treatment Diabetic Retinopathy Study (ETDRS) letters, reduction of central retinal thickness, and partial or complete absorption of subretinal fluid (SRF).

These guidelines underscore the significance of regular administration of intravitreal anti-VEGF medications to prevent blindness in most patients with wet AMD.

Without such treatment, patients experience severe, irreversible vision loss.

* Optical Coherence Tomography (OCT): Used for detailed imaging of retinal structure and fluid, aiding in the diagnosis and monitoring of wet AMD.
* Anti-VEGF Treatments: Recommended for late AMD (wet active) to prevent vision loss.
* Depression Monitoring: Important due to the increased risk in patients with AMD.
* Low-Vision Services: Considered for patients with AMD causing visual impairment.
* Group-Based Rehabilitation Programs: Promote independent living for people with AMD.

These guidelines aim to enhance patient care and improve outcomes for individuals with wet AMD by emphasizing timely intervention and comprehensive management strategies

**2. DRY (ATROPHIC) MACULAR DEGENERATION**

Dry macular degeneration is an eye condition that causes blurred vision or reduced central vision. It is caused by the breakdown of a part of the retina known as the macula (MAK-u-luh). The macula is responsible for central vision. This condition is common among people over 50.

Dry macular degeneration may start in one eye then develop in the other eye. It also may develop in both eyes at the same time. Over time, vision may worsen and affect the ability to do things, such as read, drive and recognize faces. But having dry macular degeneration doesn't mean you'll lose all your sight. Vision loss is typically central, and people retain their side vision. Some people have only mild central vision loss. In others, it can be more severe.

This form is quite common. About 80 percent (8 out of 10) of people who have AMD have the dry form. Dry AMD is when parts of the macula get thinner with age and tiny clumps of protein called drusen grow. People with dry AMD may have drusen, pigment abnormalities, or geographic atrophy (an area of cell loss in the retina). You slowly lose central vision.

Early detection and self-care measures may delay vision loss caused by dry macular degeneration.

**CAUSES**

No one knows exactly what causes dry macular degeneration. Research suggests that it may be a combination of genes and other factors, including smoking, obesity and diet.

The condition develops as the eye ages. Dry macular degeneration affects the macula. The macula is the area of the retina that's responsible for clear vision in the direct line of sight. Over time, tissue in the macula may thin and lose cells responsible for vision.

**RISK FACTORS**

Factors that may increase the risk of macular degeneration include:

* **Age.** This disease is most common in people over 50.
* **Family history and genetics.** This disease has a hereditary component, meaning it runs in families. Researchers have identified several genes linked to the condition.
* **Race.** Macular degeneration is more common in white people.
* **Smoking.** Smoking cigarettes or being exposed to tobacco smoke on a regular basis greatly increases the risk of macular degeneration.
* **Obesity.** Research suggests that obesity may increase the chance that early or intermediate macular degeneration will progress to the more serious form of the disease.
* **Cardiovascular disease.** If you have heart or blood vessel disease, called cardiovascular disease, you may be at higher risk of macular degeneration.

**SYMPTOMS**

Dry macular degeneration symptoms usually develop gradually and without pain. They may include:

* Visual distortions, such as straight lines seeming bent.
* Reduced central vision in one or both eyes.
* The need for brighter light when reading or doing close-up work.
* Increased difficulty adapting to low light levels, such as when entering a dimly lit restaurant or theater.
* Increased blurriness of printed words.
* Difficulty recognizing faces.
* A well-defined blurry spot or blind spot in the field of vision.

Dry macular degeneration can affect one or both eyes. If only one eye is affected, you may not notice any changes in your vision. This is because your good eye may compensate for the affected eye. And the condition doesn't affect the side vision, so it does not cause total blindness.

Dry macular degeneration is one of two types of age-related macular degeneration. It can progress to wet macular degeneration, which is when blood vessels grow and leak under the retina. The dry type is more common, but it usually progresses slowly over years. The wet type is more likely to cause a relatively sudden change in vision resulting in serious vision loss.

**DIAGNOSIS METHODS**

An eye care professional may diagnose dry macular degeneration by reviewing medical and family history and doing a complete eye exam. Other tests may be done, including:

* **Examination of the back of the eye.** An eye doctor puts drops in the eyes to dilate them and uses a special tool to examine the back of the eye. The eye doctor looks for a mottled appearance that's caused by yellow deposits that form under the retina, called drusen. People with macular degeneration often have many drusen.
* **A test for changes in the center of the vision field.** An Amsler grid may be used to test for changes in the center of the vision field. If you have macular degeneration, some of the straight lines in the grid may look faded, broken or distorted.
* **Fluorescein angiography.** During this test, an eye doctor injects a dye into a vein in the arm. The dye travels to and highlights the blood vessels in the eye. A special camera takes pictures as the dye travels through the blood vessels. The images may show retinal or blood vessel changes.
* **Indocyanine green angiography.** Like fluorescein angiography, this test uses an injected dye. It may be used alongside a fluorescein angiogram to identify specific types of macular degeneration.
* **Optical coherence tomography.** This noninvasive imaging test displays detailed cross sections of the retina. It identifies areas of thinning, thickening or swelling. These can be caused by fluid buildup from leaking blood vessels in and under the retina.

**TREATMENT OPTIONS**

For now, there's no way to reverse damage from dry macular degeneration. However, there are many clinical trials in progress. If the condition is diagnosed early, you can take steps to help slow its progression, such as taking vitamin supplements, eating healthy and not smoking.

**Vitamin supplements**

For people with intermediate or advanced disease, taking a high-dose formulation of antioxidant vitamins and minerals may help reduce the risk of vision loss. Research from the Age-Related Eye Disease Study 2 (AREDS2) has shown benefit in a formulation that includes:

* 500 milligrams (mg) of vitamin C.
* 400 international units (IU) of vitamin E.
* 10 mg of lutein.
* 2 mg of zeaxanthin.
* 80 mg of zinc as zinc oxide.
* 2 mg of copper as cupric oxide.

The evidence doesn't show benefit in taking these supplements for people with early-stage dry macular degeneration. Ask your eye doctor if taking supplements is right for you.

**Low vision rehabilitation**

Age-related macular degeneration doesn't affect your side vision and typically doesn't cause total blindness. But it can reduce or eliminate central vision. You need central vision to read, drive and recognize people's faces. It may help for you to get care from a low vision rehabilitation specialist, an occupational therapist, your eye doctor and others trained in low vision rehabilitation. They can help you find ways to adapt to your changing vision.

**Surgery to implant a telescopic lens**

For some people with advanced dry macular degeneration in both eyes, an option to improve vision may be surgery to implant a telescopic lens in one eye. The telescopic lens, which looks like a tiny plastic tube, has lenses that magnify your field of vision. The telescopic lens implant may improve both distance and close-up vision, but it has a very narrow field of view. It can be useful in urban settings as an aid to see street signs.

**Lifestyle and home remedies**

Even after receiving a diagnosis of dry macular degeneration, these steps may help slow vision loss.

* **Don't smoke.** If you smoke, ask a healthcare professional for help quitting.
* **Choose a healthy diet.** The antioxidant vitamins in fruits and vegetables contribute to eye health. Kale, spinach, broccoli, squash and other vegetables have high levels of antioxidants, including lutein and zeaxanthin. These nutrients may benefit people with macular degeneration.

Eating foods with high levels of zinc also may be helpful for people with macular degeneration. These include high-protein foods, such as beef, pork and lamb. Non-meat sources include milk, cheese, yogurt, whole-grain cereals and whole-wheat bread.

Another good choice is healthy unsaturated fat, such as in olive oil. And research studies have shown that a diet high in omega-3 fatty acids, such as in salmon, tuna and walnuts, may lower the risk of advanced age-related macular degeneration. But the same benefit is not shown from taking omega-3 supplements, such as fish oil pills.

* **Manage your other medical conditions.** If you have cardiovascular disease or high blood pressure, for example, take your medicine and follow your healthcare team's instructions for controlling the condition.
* **Maintain a healthy weight and exercise regularly.** If you need to lose weight, reduce the number of calories you eat and increase the amount of exercise you get each day.
* **Have routine eye exams.** Ask your eye doctor about the recommended schedule for follow-up exams. In between checkups, you can do a self-assessment of your vision using an Amsler grid. These steps will help tell you if your condition develops into wet macular degeneration, which can be treated with medicines.

**Should you take nutritional supplements for AMD?**

Talk with your ophthalmologist about whether nutritional supplements are recommended for you. Here are some topics to discuss:

* **Your chance of getting advanced AMD.** Studies show that nutritional supplements might help people with early to intermediate AMD who are at risk for developing advanced AMD.
* **Eye-healthy foods.** Studies show that nutritional supplements alone are not enough to prevent or delay advanced AMD. You also should eat a healthy, balanced diet. This includes dark leafy greens (like spinach and kale) along with yellow, orange and other colorful fruits and vegetables. Eating fatty fish like salmon may also lower your risk of early or advanced AMD.
* **Benefits and risks of nutritional supplements.** Taking nutritional supplements can be helpful, but there can be possible health risks. For example, a type of [vitamin B3 called nicotinamide](https://www.aao.org/eye-health/tips-prevention/vitamin-b3-nicotinamide-glaucoma) is being studied for its potential benefits for people with glaucoma but it may also cause severe liver injury. Talk with your ophthalmologist and primary care doctor about how the vitamins and minerals listed above might affect you.

**PREVENTION TIPS**

It's important to have routine eye exams to identify early signs of macular degeneration. The following measures may help reduce the risk of developing dry macular degeneration:

* **Manage all medical conditions.** For example, if you have cardiovascular disease or high blood pressure, take your medicine and follow your healthcare team's instructions for controlling the condition.
* **Don't smoke.** People who smoke are more likely to develop macular degeneration than are people who don't smoke. Ask a healthcare professional for help stopping smoking.
* **Maintain a healthy weight and exercise regularly.** If you need to lose weight, reduce the number of calories you eat and increase the amount of exercise you get each day.
* **Choose a diet rich in fruits and vegetables.** These foods contain antioxidant vitamins that reduce your risk of developing macular degeneration.
* **Include fish in your diet.** Omega-3 fatty acids, which are found in fish, may reduce the risk of macular degeneration. Nuts such as walnuts also contain omega-3 fatty acids

**POSSIBLE COMPLICATIONS**

People whose dry macular degeneration has progressed to central vision loss have a higher risk of depression and social isolation. With profound loss of vision, people may see visual hallucinations. This condition is called Charles Bonnet syndrome. Dry macular degeneration may progress to wet macular degeneration, which can quickly cause complete vision loss if left untreated.

**WHEN TO SEE A DOCTOR / RED FLAG**

See your eye care professional if:

* You notice changes, such as distortion or blind spots, in your central vision.
* You lose the ability to see fine detail.

These changes may be the first sign of macular degeneration, particularly if you're over age 60.

With AMD you lose your central vision. You cannot see fine details, whether you are looking at something close or far. But your peripheral (side) vision will still be normal. For instance, imagine you are looking at a clock that has hands. With AMD, you might see the clock’s numbers but not the hands.

AMD is very common. It is a leading cause of vision loss in people 50 years or older.

**DIFFERENTIAL DIAGNOSIS**

Reticular pseudodrusen should be differentiated from drusen. The former are deposits above the RPE that are in the subretinal plane.

Fundus features of dry ARMD should also be differentiated from adult vitelliform macular dystrophy and retinal drug toxicity. Fundus in adult vitelliform dystrophy reveals yellowish subretinal deposits, whereas RPE pigmentary changes due to drugs like hydroxychloroquine, deferoxamine, and cisplatin can mimic the scene in Dry ARMD.

Neovascular AMD should be differentiated from retinal angiomatous proliferation and polypoidal choroidal vasculopathy (PCV). Retinal angiomatous proliferation is also known as type 3 CNVM, where the neovascularization begins within the retina and then progresses towards the RPE and choroid. On FFA, early phases reveal a retinal vessel dipping perpendicularly into the CNVM component, indicating the retinochoroidal anastomosis.

Polypoidal choroidal vasculopathy can be differentiated on fundus examination by the presence of orangish nodules along with subretinal hemorrhage or fluid. OCT shows large choroidal vessels, also known as pachy vessels, in these cases. The choroid is thickened, unlike ARMD, where the choroid is thin. ICGA shows polyps that are diagnostic of PCV.

**RECOMMENDATION**

Here are some important points to remember:

* ARMD is the leading cause of blindness in the elderly age group.
* Regular follow up in cases of early ARMD can help in the timely recognition of signs of progression and facilitate initiation of treatment.
* With the advent of SD-OCT, the follow up of these patients has improved significantly. Various prognostic markers that have been identified on SD-OCT are useful clinically in decision making regarding treatment and patient education and counseling regarding the visual prognosis.
* Intravitreal anti-VEGF injections have largely replaced all the other treatment modalities. The visual acuity achieved at the end of the treatment period is much better than that achieved with the laser. Patients' quality of life has improved. With treat and extend regimen being used widely, the burden of the cost of injections, as well as the number of visits to the hospital, have reduced significantly.
* Low vision aids are very helpful in rehabilitating the patients who progress to blindness despite treatment.
* The disease is a bilateral process that makes the patient dependent and affects the quality of life.
* The cost of intravitreal injections is a burden for the patient.
* Regular follow up in cases of monthly dosing also increases the number of visits to the hospital.

**EPIDEMIOLOGY**

The disease is estimated to affect around 196 million people by 2020 and 288 million people by 2040. Early AMD is more common in individuals of European ancestry than in Asians, whereas the prevalence of late AMD is the same between the two populations. In 2015, AMD was the fourth most common cause of blindness globally and the third most common cause for moderate to severe vision loss. This shows the increasing importance of AMD globally.

**EPIDEMIOLOGY OF MACULAR DEGENERATION**

**How common is macular degeneration?**

Almost 20 million U.S. adults have macular degeneration. Globally, the prediction is that 288 million people will have the condition by 2040.

In the U.S., macular degeneration is a leading cause of vision loss in people who are 60 and older.

**Who might get macular degeneration?**

As the term “age-related macular degeneration” (AMD) implies, macular degeneration is more likely to occur as you get older. However, people can develop macular degeneration at younger ages because of several factors.

In addition to age, risk factors for macular degeneration include:

* Having a family history of macular degeneration.
* Being overweight.
* Smoking.
* Having high blood pressure (hypertension).
* Eating a diet high in saturated fats.
* Being white.

**POSSIBLE COMPLICATIONS**

**What are the complications of macular degeneration?**

Losing your central vision can make it challenging to do certain tasks. Depending on the extent of vision impairment, you may not be able to:

* Read well.
* Recognize faces.
* Drive.
* Cook.
* Do home repairs.

Severe AMD may lead to you being legally blind.

Changes in your lifestyle can lead to depression and anxiety. Some people with AMD experience Charles Bonnet syndrome, a condition that causes visual hallucinations.

REFERENCES: <https://my.clevelandclinic.org/health/diseases/15246-macular-degeneration>

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[Dry macular degeneration - Symptoms and causes - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/dry-macular-degeneration/symptoms-causes/syc-20350375)

[Macular Degeneration - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK560778/)

**ALBINISM**

*ALTERNATIVE NAMES:* Albinism is also known as “achromia”, “achromasia”, and “achromatosis”

**DEFINITION / DESCRIPTION**

Albinism is a genetic disorder where you’re born with less melanin pigment than usual. Melanin is a chemical in your body that determines the color of your skin, hair and eyes. It’s also involved in optic nerve development, which means it helps your eyes function as they should.

Most people with albinism have very pale skin, hair and eyes. The exact skin tone, hair color and eye color can vary from person to person. Most people with this condition also have vision problems ranging from mild to severe.

The word “albino” comes from the Latin word “albus,” which means white. You might hear someone use “albino” to refer to a person with albinism. But healthcare providers and many people with this condition prefer to use “a person with albinism.” This term puts the person first rather than using a medical condition to define their identity.

#### **Types of albinism**

There are two main types of albinism:

* Oculocutaneous albinism (OCA): Oculocutaneous (pronounced “ock-you-low-kew-TAIN-ee-us”) albinism is the most common type of albinism. People with OCA have extremely pale hair, skin and eyes. They typically also have vision problems. There are seven forms of OCA, and each affects your body in a slightly different way. For example, your hair and skin may be lighter or darker depending on the specific form of OCA you have.
* Ocular albinism (OA): Ocular albinism is much less common than OCA. Ocular albinism mostly affects your eyes. It doesn’t affect your skin or hair much, if at all. OA usually leads to blurred vision, sensitivity to light and other symptoms that may affect how you see and interact with the world around you.

Albinism is sometimes a feature of a genetic syndrome. This means you have OCA or OA, as well as other medical conditions affecting different parts of your body. For example, albinism occurs as part of:

* Hermansky-Pudlak syndrome (HPS): This is a genetic metabolic disorder. People with HPS have albinism, as well as blood disorders, bruising issues and lung, kidney or bowel diseases.
* Chediak-Higashi syndrome (CHS): This is a genetic immune disorder that makes you more vulnerable to infections. People with CHS also commonly have albinism and may bruise or bleed more easily than expected.

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Albinism can affect people of all races and ethnic groups, including Black people. Researchers estimate that OCA affects 1 in 20,000 people around the world. OA affects at least 1 in 60,000 males.

**CAUSES**

Variations in the genes responsible for melanin production cause albinism. Specific genes associated with oculocutaneous albinism include:

* *TYR.*
* *OCA2.*
* *TYRP1.*
* *SLC45A2.*

Variations in the *GPR143* gene are associated with ocular albinism.

Some people with albinism don’t have variations in any of these genes. In these cases, the exact genetic cause is unknown.

#### **Is albinism genetic?**

Yes, albinism is passed down (inherited) through families.

Oculocutaneous albinism (OCA) follows an autosomal recessive pattern of inheritance. This means you must inherit an albinism gene from both of your biological parents to develop the condition yourself.

If just one of your parents has an albinism gene, you won’t be born with OCA. But you’ll have a 50% chance of being a carrier of the gene. If you’re a carrier, that means you have one albinism gene but don’t show any signs or symptoms of the condition. If you have a baby with a person who’s also a carrier, your baby will have a 25% chance of being born with OCA.

Ocular albinism (OA) usually follows an X-linked pattern of inheritance. This means the genetic variation is passed through the X chromosome. OA mostly occurs in males.

#### **Is albinism a disease?**

Albinism isn’t a disease. Albinism is a genetic condition that people are born with. It’s not contagious and it can’t be spread.

**RISK FACTORS**

Albinism is an inherited genetic disorder. Usually, both parents must carry the albinism gene to have a child with albinism. The albinism gene is a recessive gene, meaning that a child has to receive a copy from both parents to have the disorder. If the child gets a copy of the gene from just one parent, he or she will not have symptoms of albinism. If both parents carry the gene, there is a one-in-four chance with each pregnancy that the baby will be born with albinism.

One type of albinism, called X-linked ocular albinism, is usually inherited from the mother. In this case, the gene for albinism is located on an X chromosome. Females have two X chromosomes, while males have one X chromosome and one Y chromosome. X-linked ocular albinism appears almost exclusively in males. The gene for it is passed from mothers (who carry it without developing the condition) to their sons. The mothers generally have normal vision. For each son born to a mother who carries the gene, there is a one-in-two chance of having X-linked ocular albinism.

**SIGNS / SYMPTOMS (Albinism symptoms**)

Albinism typically affects the appearance of your skin, hair and eyes. It may also affect your vision, or how you see the world around you.

#### **Skin**

People with albinism often have very pale skin. But your exact skin tone may be lighter or darker depending on the type of albinism you have and how much melanin your body produces.

For example, if you have ocular albinism (OA), your skin tone may be affected very little or not at all. It may resemble that of your biological parents and siblings or look just a little bit lighter.

With oculocutaneous albinism (OCA), your skin tone depends on the specific form of the condition. Here are a few examples:

* Type 1 OCA: Very pale skin.
* Types 2 and 4 OCA: Creamy white skin.
* Type 3 OCA: Reddish-brown skin. In general, most people with albinism have a low amount of melanin in their skin (hypopigmentation). This means you’ll get sunburnt more quickly than others when outdoors, and you’re at a higher risk of developing skin cancer.

#### **Hair**

Hair color also varies by albinism type. People with OCA type 1 often have white hair, while those with other types may have hair that’s light yellow, blond, light brown or red. It all depends on how much melanin your body produces. The less melanin you have, the lighter your hair will be.

#### **Eyes**

Many people have blue eyes (often very pale). Others have hazel or brown eyes. But albinism doesn’t just affect eye color. It also affects how your eyes develop and function.

People with albinism can experience a wide range of eye-related signs and symptoms, including:

* Blurry or distorted vision.
* Refractive errors.
* Reduced depth perception.
* Crossed eyes (strabismus).
* Rapid eye movements (nystagmus).
* Light sensitivity (photophobia).

**DIAGNOSIS METHODS**

To diagnose albinism, an ophthalmologist will give you a thorough eye examination. He or she will look for nystagmus, strabismus and photophobia. Any one of these conditions by itself is not necessarily a sign of albinism. An ophthalmologist will also look at the retina and iris to see if they have developed as they should.

**ALBINISM TREATMENT OPTIONS**

Albinism itself has no treatment. But some conditions that people with albinism have are treatable. Other conditions related to albinism are manageable.

For example, strabismus can be treated with glasses or surgery. Glasses can also help improve vision and reduce light sensitivity. For children with low vision, low vision aids such as hand-held magnifiers can help. Glasses with small telescopes attached are helpful for older children and adults. These lenses can help with both close and distant vision.

Parents and teachers can work together to help a child with albinism. It's important to consider seating, lighting and optical aids in the classroom. These can make learning easier for a child with albinism. Some students with albinism may benefit from having a teacher for the visually impaired (or TVI).

Peer support groups can help children and adults with albinism. These groups can help the individual to:

* feel less isolated
* learn positive attitudes and coping skills from others with low vision;
* gather valuable resource information.

**Prevention tips**

Because albinism is inherited, there’s no way to prevent it. But if you have a family history of albinism and are planning to have children, you might want to consider genetic counseling.

**OUTLOOK / PROGNOSIS**

Life expectancy within the non-syndromic OCA population is comparable to the general population. There is an increased mortality risk due to skin cancer. This risk changes based on the amount of relative sun exposure in a geographic area and certain socio economic issues. These socioeconomic issues include limited access to sunscreen, limited education on sun-protective measures, cultural differences in dress, limited access to healthcare professionals for surveillance leading to late presentation and late treatment, inability to comply with or complete treatment courses. In these same lower socioeconomic regions, there is often a palpable stigma associated with albinism, and the afflicted may be victims of persecution, prejudice, and violence. Some albinos have even been murdered as their organs are highly valued on the black market. Albinos have normal intelligence compared with the general population. There is some delayed visual maturation, and this can lead to educational delay if not addressed early enough. Furthermore, poor self-image and social alienation can lead to feelings of isolation and depression. Albinos do have an increased rate of attention deficit disorder.

**POSSIBLE COMPLICATION**

Visual deficits can lead to the following:

* Limited work opportunities where a minimum visual acuity is required
* Difficulty in reading due to uncorrected visual deficits that may lead to educational delays
* Inability to obtain driver’s license due to visual impairment

Infection risk in Chediak-Higashi syndrome

Bleeding Diathesis in Hermansky-Pudlak syndrome

Solar damage: Pachydermia (coarse, rough, thick skin), actinic keratoses, solar lentigines (in non-OCA1A albinos), solar erythema

Malignancy: Basal and squamous cell carcinomas (BCC, SCC). SCC is by far the most common malignancy reported among albinos (representing more than 75% of cases of malignancy), developing as early as the teens. One study found that more than 70% of them were diagnosed in the head and neck areas. Another study found the head, face, and hands equally affected at approximately 20% each. Albinism increases the relative risk of SCC as much as 1000 times, at least in the African population. Cumulative sun exposure is the major risk factor, with the incidence of cancers increasing with patient age. BCC accounts for about 24% of malignancies, with the remaining 1% being melanoma. Melanomas are rare in albinos with only a few case reports in the literature, despite melanocytes still being present in the skin in the same relative number. Surprisingly, melanomas often occur in non-exposed areas, which speaks against UV-radiation being the culprit. Practitioners need to have a high level of suspicion since these lesions are both rare and tend to be amelanotic in albinos.

**When to see a doctor / red flag**

Your healthcare providers will tell you how often you should come in for appointments. Be sure to follow the schedule they provide.

Call a provider if:

* Your vision seems poorer or less clear than usual.
* You have any new or changing eye-related symptoms.
* You notice any skin changes.

**DIFFERENTIAL DIAGNOSIS**

Based upon similar ocular findings (early onset nystagmus):

* OCA syndromes
* Ocular albinism
* Optic nerve atrophy and hypoplasia: The condition of broad etiology resulting in decreased visual acuity, nystagmus, and optic nerve atrophy, optic disc pallor
* Inherited retinal dystrophy: A family of disorders of variable heritance patterns (autosomal dominant, X-linked, recessive) and broad phenotypes that included congenital nystagmus and decreased visual acuity
* FRMD7-related infantile nystagmus syndrome (also known as congenital motor nystagmus, congenital infantile nystagmus): An X-linked disorder with nystagmus and reduced visual acuity with normal VEPs
* Aniridia: The absence of the iris, which demonstrates foveal hypoplasia, nystagmus, amblyopia, and cataracts
* Aland Island eye disease (Forsius-Eriksson syndrome): An X-linked disorder with fundus hypopigmentation, foveal hypoplasia, myopia, nystagmus, astigmatism, night blindness
* Cross-McKusick-Breen syndrome: An autosomal recessive disorder with hypopigmentation of skin, silvery gray hair, microphthalmia, corneal opacification, nystagmus, with or without mental retardation, athetosis, ataxia, joint contractures, and spastic tetraplegia
* Achromatopsia: An autosomal recessive disorder causing dysfunctional cone cells in the retina, resulting in partial or complete loss of color vision with associated photophobia, nystagmus, low visual acuity, and hyperopia.

Based on hair and cutaneous finding (hypopigmentation):

* OCA syndromes
* Hermansky-Pudlak syndrome
* Chediak-Higashi syndrome
* Angelman syndrome and Prader-Willi syndrome
* Vici syndrome: An autosomal recessive disorder with absent corpus callosum; subjects present with hair and skin hypopigmentation, findings similar to OA1, microcephaly, immunodeficiency, cardiac abnormalities, failure to thrive, cataracts, cleft lip and palate, and neurologic abnormalities
* Waardenburg syndrome type II: Autosomal dominant MITF gene mutation presenting with patchy skin hypopigmentation, white forelock or prematurely gray hair, iris heterochromia, sensorineural hearing loss
* Tietz albinism-deafness syndrome: Autosomal dominant *MITF* gene mutation presenting with white eyebrows and eyelashes, iris hypopigmentation, normal visual acuity, and sensorineural hearing loss
* Griscelli syndrome: Autosomal recessive defects in myosin, myosin receptors, and binding; melanocytes fail to transfer melanosomes to dendrites and peripheral keratinocytes leading to diluted skin/hair color; present with hypopigmentation, melanin aggregation in silvery-gray hair, immunodeficiency, decreased visual acuity with roving eye movements, pancytopenia, hemophagocytic syndrome, and cerebral demyelination.

**RECENT GUIDELINES OR UPDATES**

Due to the heterogeneity of symptoms and signs of OCA and OA, a multidisciplinary team approach involving Ophthalmologist and Dermatologist is required in assessing and managing patients with albinism. Children with albinism should undergo cycloplegic refraction to ensure prompt correction of refractive error. Careful assessment of nystagmus is also essential, noting its plane, axis of oscillation, conjugacy, frequency and amplitude. Modern video-based eye movement recordings can help in analyzing the nystagmus waveform and characteristics. AHP when the null zone is not in the primary position can be objectively measured using torticollometer, Harms’ wall and orthopedic goniometer. Accurate measurement of deviation using prism cover tests is also crucial as patients with albinism often have strabismus. Other investigations include VEPs, which would show misrouting of visual pathways with abnormally high numbers of temporal fibers decussating at the optic chiasm. The International Society for Clinical Electrophysiology of Vision standard recommended multichannel ‘onset-offset’ pattern VEP presented with a field of 30 degrees to record chiasmal misrouting. OCT is another valuable tool to assess the degree of foveal hypoplasia, which is graded using the Leicester Grading System

Differentiating subtypes of albinism based on clinical presentation alone is challenging due to overlapping clinical features. Thus, genetic testing is helpful in confirming clinical suspicion, obtaining an accurate diagnosis and can aid in discussing prognosis and genetic counseling. This includes the differences in prognosis between syndromic forms compared to non-syndromic forms and searching for systemic phenotypes in syndromic OCA. Prior to genetic testing, a detailed family history should be taken, with attention to relatives with clinical manifestations of OCA and OA. Molecular genetic testing approaches can include a combination of targeted testing (multigene panel and chromosomal microarray) and comprehensive genomic testing (exome sequencing or genome sequencing). The genetic information on causative pathologic variants is important for carrier testing of family members, prenatal diagnosis and genetic counseling. Issues related to genetic counseling that need to be explored further include family planning, DNA banking and prenatal testing

One of the challenges in obtaining a genetic diagnosis of albinism has been due to missing heritability or incomplete genetic diagnosis (where only one pathogenic *TYR* variant is identified). However, work by Gronskov *et al*. highlighted that a pathogenic haplotype (the allele p.S192Y-p.R402Q in *cis*, (also referred to as ‘cis-YQ’)) is a significant contributor to the disease and is common in Europeans. They suggest that these variants in combination may have an additive effect. This has been verified by other groups. Michaud *et al*. suggested that a promoter variant had a significant role in the pathogenesis of *TYR* associated albinism, however a recent study by Loftus *et al*. did not find a difference in *TYR* mRNA expression in primary melanocytes between the different promoter genotypes. To identify the pathogenic haplotype, phasing of *TYR* variants is recommended. This is a scenario where long-read sequencing could help resolve whether the pathogenic haplotype is present. If there is still missing heritability, investigating for structural variants is recommended recommended.ly in carriers with albinism, phenotypic manifestations have also been reported.

Recent advances in genetics offer the possibility of precise diagnosis. This includes the application of adeno-associated virus-based gene therapy for OA1, which showed positive outcomes in animal models. Gene editing of *TYR* gene with the CRISPR-associated nuclease protein (CRISPR/Cas9) system was also demonstrated on animal level by Song *et al.* More recently, a patient-derived stem cell model for studying eye conditions related to OCA was developed. This is believed to be an important step forward in understanding albinism and testing potential therapies to treat it. Investigating albinism in some animal models may be limited as they lack fovea.

Management of albinism at present usually includes supportive treatment aimed at optimizing vision, managing and addressing clinical manifestations especially photophobia, protecting retina from ultraviolet rays and reducing related complications such as skin cancers. Correction of refractive error using spectacles and/or contact lenses to reduce the risk of amblyopia is essential. When considering the choice of contact lenses, soft contact lenses were associated with worse visual acuity compared to rigid gas permeable lenses and spectacles in a randomized controlled trial. Patients with albinism may have nystagmus which could cause misalignment of the toric soft contact lenses resulting in blurred vision. Some patients with albinism also do not find refractive correction helpful as their vision was limited by foveal hypoplasia. In cases of poor vision, low visual aids are helpful. Nystagmus, strabismus and AHP can be managed by prism glasses or surgery. The Anderson-Kestenbaum procedure is an effective surgical approach to move null points to correct AHP. However, there may be a higher risk of non-optimal success after surgery among patients with albinism due to poor fusion secondary to lower visual acuity and nystagmus . Tinted glasses or contact lenses can help to improve glare . Individuals with albinism should be advised to wear sun protective clothing and apply sunscreens outdoors. Psychosocial and educational support should also be provided to individuals with albinism

**EPIDEMIOLOGY**

### Frequency

The incidence of albinism is 1 in 20,000 persons worldwide, compared with a rate of 1 in 37,000 in the United States. OCA1 is the most commonly found subtype in Caucasians, and accounting for 50% of all cases worldwide. OCA2 is responsible for 30% of cases worldwide and is more common in Africa. OCA3 affects 3% and OCA4 affects 17% of all cases globally.6 OCA5-8 are rare forms. HPS is a common type of albinism in Puerto Rico, but the disorder is rare in other parts of the world

Mortality/Morbidity

Albinism usually is not linked to mortality, and individuals with the disorder have a normal lifespan; the overall health of children and adults with albinism usually does not suffer from the decreased melanin in the hair, skin, and eyes, and this reduction causes no additional systemic effects. [7]

Normal growth and intellectual development should progress in a child with albinism, and they should accomplish developmental milestones on par with other children their age.

The bulk of morbidity linked to albinism is related to visual impairment, photosensitivity of the skin, and increased risk for cutaneous cancer.

Those with syndromes related to albinism, such as HPS or CHS, may experience hearing impairment or abnormal blood clotting.

*REFERENCE:*

[Full article: Our current understanding of clinical characteristics and the genetics of patients with albinism](https://www.tandfonline.com/doi/full/10.1080/17469899.2024.2320117#d1e940)

[Albinism - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK519018/)

**ALZHEIMER’S DISEASE**

Another name for Alzheimer’s disease is “dementia and the eye”.

**DEFINITION / DESCRIPTION**

Alzheimer's disease is the most common cause of dementia. Alzheimer's disease is the biological process that begins with the appearance of a buildup of proteins in the form of amyloid plaques and neurofibrillary tangles in the brain. This causes brain cells to die over time and the brain to shrink.

About 6.9 million people in the United States age 65 and older live with Alzheimer's disease. Among them, more than 70% are age 75 and older. Of the more than 55 million people in the world with dementia, 60% to 70% are estimated to have Alzheimer's disease.

Early symptoms of Alzheimer's disease include forgetting recent events or conversations. Over time, Alzheimer's disease leads to serious memory loss and affects a person's ability to do everyday tasks.

There is no cure for Alzheimer's disease. In advanced stages, loss of brain function can cause dehydration, poor nutrition or infection. These complications can result in death.

But medicines may improve symptoms or slow the decline in thinking. Programs and services can help support people with the disease and their caregivers.

**CAUSES AND RISK FACTORS**

The exact causes of Alzheimer's disease aren't fully understood. But at a basic level, brain proteins don't function as usual. This disrupts the work of brain cells, also known as neurons, and triggers a series of events. The neurons become damaged and lose connections to each other. They eventually die.

Scientists believe that for most people, Alzheimer's disease is caused by a combination of genetic, lifestyle and environmental factors that affect the brain over time. In less than 1% of people, Alzheimer's is caused by specific genetic changes that almost guarantee a person will develop the disease. For people in this group, the disease usually begins in middle age.

The disease begins years before the first symptoms. The damage most often starts in the region of the brain that manages memory. The loss of neurons spreads in a somewhat predictable pattern to other regions of the brain. By the late stage of the disease, the brain has shrunk.

Researchers trying to understand the cause of Alzheimer's disease are focused on the role of two proteins:

* **Plaques.** Beta-amyloid is a fragment of a larger protein. When these fragments clump together, they affect communication between brain cells. The clumps form larger deposits called amyloid plaques.
* **Tangles.** Tau proteins play a part in a brain cell's internal support and transport system to carry nutrients and other essential materials. In Alzheimer's disease, tau proteins change mshape and organize into structures called neurofibrillary tangles. The tangles disrupt the transport system and cause damage to cells.

**Causes and risk factors you can modify**

**Head trauma**

People who have experienced serious head injuries are at higher risk of developing Alzheimer’s. The risk increases if the injury involves losing consciousness or happens repeatedly, such as in contact sports.

By wearing a helmet during contact sports such as football and hockey or avoiding these activities altogether, you may be able to reduce your chances of experiencing this type of injury.

**Smoking**

Researchers have identified smoking as a risk factor for Alzheimer’s. A 2023 study from Korea found that quitting smoking was associated with an 8% lower risk of all types of dementia and a 6% lower risk of Alzheimer’s specifically.

**High blood pressure**

High blood pressure may increase your risk of developing Alzheimer’s. According to a 2010 research review, having high blood pressure in middle age is associated with a greater chance of developing Alzheimer’s than having high blood pressure later in life.

**Obesity**

In a study with more than 10,000 participants that was published in 2007, researchers found that a body mass index (BMI) of 25 or greater (overweight) was associated with double the risk of developing Alzheimer’s, and a BMI of 30 or greater (obesity) was associated with triple the risk.

**Lack of physical activity**

A low level of physical activity has been linked to dementia and Alzheimer’s disease.

While there is no specific exercise regimen that can prevent dementia and Alzheimer’s, many types of physical activity and exercise may benefit brain health. They found that any activity was helpful.

The following are some types of exercise that may help prevent Alzheimer’s disease:

* aerobic exercise, such as:
  + walking
  + running
  + dancing
  + biking
  + swimming
* muscle development exercises, such as:
  + weightlifting
  + use of gym equipment
  + dumbbell exercises
* body conditioning exercises, such as:
  + situps
  + pushups
  + lunges
  + squats

**Lack of mental activity**

Mental activity might be as important as physical activity for decreasing your risk of Alzheimer’s disease. Examples of mental activities include:

* taking a class
* socializing with family and friends
* volunteering in your community
* playing board games or cards
* reading

These mental activities may help maintain your cognitive (thinking) ability. Social interaction also helps. The key is to pick activities that challenge you.

Researchers are not sure why this works. One theory is that these challenges help your brain develop more internal connections, which protect against dementia.

**Diet**

A heart-healthy diet may benefit cognitive function, according to the Alzheimer’s Association. The Dietary Approaches to Stop Hypertension (DASH) diet and the Mediterranean diet may lower your risk of both heart disease and dementia.

These diets involve:

* eating lots of fruits, vegetables, and low fat dairy
* eating poultry, fish, and whole grains
* eating foods that are low in saturated fat, total fat, and cholesterol
* limiting red meat, sweets, sugary beverages, and sodium

**Causes and risk factors you cannot modify**

**Age**

Alzheimer’s is not a natural part of growing older, but older age is a risk factor for developing it. According to the Alzheimer’s Association, about [1 in 9 people](https://www.alz.org/alzheimers-dementia/facts-figures) ages 65 and older in the United States have Alzheimer’s, and 73% of them are 75 or older.

**Gender**

Women outnumber men when it comes to Alzheimer’s. In fact, almost two-thirds of people living with Alzheimer’s in the United States are women.

By the time they reach 65 years of age, women have a 20% chance of developing Alzheimer’s. Women in their 60s are twice as likely to develop Alzheimer’s as they are to get breast cancer.

One of the main reasons women may have higher rates of Alzheimer’s than men is that women tend to live longer, on average, and older age is a significant risk factor for the condition. But researchers have started to explore whether women may be at a higher risk for Alzheimer’s because of biological or genetic differences, regardless of age.

**Genes**

Researchers have found two classes of genes related to Alzheimer’s: deterministic genes and risk genes.

Deterministic genes nearly guarantee that people will develop the disease if they live long enough. People with deterministic genes often develop symptoms of Alzheimer’s sometime in their early 40s through their mid-50s.

But this is rare: Deterministic genes cause about 1% of all Alzheimer’s cases.

People with risk genes may or may not develop the disease, but they’re more likely to develop it than people without risk genes. The gene that’s most commonly associated with Alzheimer’s is called apolipoprotein E-e4 (APOE-e4).

**Family history**

Alzheimer’s often [runs in families](https://www.alz.org/alzheimers-dementia/what-is-alzheimers/causes-and-risk-factors). If you have a parent, sibling, or child with the disease, you’re more likely to develop it yourself. Your risk goes up if multiple family members have Alzheimer’s. This could be due to genes, lifestyle factors, or a combination of the two.

The gene APOE-e4 plays a role here too. APOE-e4 coupled with a family history of the disease significantly increases your risk.

**Brain abnormalities**

Scientists have identified brain abnormalities in people who are likely to later develop Alzheimer’s. One is the presence of tiny clumps of protein known as plaques. The other is twisted protein strands or tangles. Inflammation, tissue shrinkage, and loss of connection between brain cells are other clues that Alzheimer’s may develop.

**Can someone avoid getting Alzheimer’s?**

It’s not possible to prevent Alzheimer’s because its exact causes are not well understood. However, eating a heart-healthy diet, exercising, engaging in mentally challenging activities, lowering your blood pressure, and protecting your head during physical activities such as contact sports may lower your risk.

**RISK FACTOR**

Risk factors for Alzheimer's disease include age, family history, lifestyle and other factors.

### **Older age**

Older age is the strongest known risk factor for Alzheimer's disease. Alzheimer's isn't a part of typical aging. But as you grow older, the chances of getting the disease goes up.

One study found that there were four new diagnoses per 1,000 people ages 65 to 74 every year. Among people ages 75 to 84, there were 32 new diagnoses per 1,000 people. For those 85 and older, there were 76 new diagnoses per 1,000 people.

### **Family history and genetics**

The risk of getting Alzheimer's disease is higher if a first-degree relative such as a parent or sibling was diagnosed with the disease. How genes among families affect the risk is largely not understood. The genetic factors are likely complex.

A better understood genetic factor is a form of the apolipoprotein E (APOE) gene. Having the form of the gene known as APOE e4 increases the risk of Alzheimer's disease. About 25% to 30% of the population carries APOE e4. But not everyone with this form of the gene develops the disease. Having two copies of APOE e4 increases the risk of Alzheimer's disease more than having one copy.

Scientists have found rare changes in three genes that virtually guarantee a person who inherits one of them will develop Alzheimer's. But these changes account for less than 1% of people with Alzheimer's disease.

### **Down syndrome**

Many people with Down syndrome develop Alzheimer's disease. This is likely related to having three copies of chromosome 21. Chromosome 21 is the gene involved in the production of the protein that leads to the creation of beta-amyloid. Beta-amyloid fragments can become plaques in the brain. Symptoms tend to appear 10 to 20 years earlier in people with Down syndrome than they do for the general population.

### **Sex assigned at birth**

Overall there are more women with the disease because they tend to live longer than men.

### **Mild cognitive impairment**

Someone with mild cognitive impairment, also called MCI, has a bigger decline in memory or other thinking skills than is usual for the person's age. But the decline doesn't prevent the person from functioning at work or socially.

However, people with MCI have a higher risk of getting dementia than are people who don't have mild cognitive impairment. When MCI affects mainly memory, the condition is more likely to progress to dementia due to Alzheimer's disease. A diagnosis of MCI offers people the chance to focus on healthy lifestyle changes and to come up with strategies to make up for memory loss. They also can schedule regular healthcare appointments to monitor symptoms.

### **Head injury**

Several large studies found that people age 50 or older who had a traumatic brain injury, also called TBI, had a higher risk of getting dementia and Alzheimer's disease. The risk is even higher in people with serious TBIs or multiple TBIs.

### **Air pollution**

Studies in animals have found that air pollution particulates can speed the breakdown of the nervous system. Human studies have found that air pollution exposure — especially from traffic exhaust and burning wood — is linked to a higher risk of dementia.

### **Heavy alcohol use**

Drinking large amounts of alcohol has long been known to cause brain changes. Several large studies and reviews found that alcohol misuse is linked to a higher risk of dementia, especially early-onset dementia.

### **Poor sleep patterns**

Research has shown that poor sleep patterns, such as trouble falling asleep or staying asleep, are linked to a raised risk of Alzheimer's disease. Sleep apnea also may raise the risk of dementia.

### **Lifestyle and heart health**

Research has shown that the same risk factors for heart disease also may increase the risk of dementia. It's not clear if these factors raise risk by worsening Alzheimer's changes in the brain or by leading to blood vessel changes in the brain. The factors include:

* Lack of exercise.
* Obesity.
* Smoking or exposure to secondhand smoke.
* High blood pressure.
* High cholesterol.
* Poorly managed type 2 diabetes.

High levels of low-density lipoprotein, known as LDL, cholesterol in middle age, in particular, raises the risk of dementia. Research has found that people younger than 65 with high LDL cholesterol levels have a higher risk of dementia. But taking medicines to lower LDL cholesterol didn't raise the risk.

These factors can all be modified, so changing lifestyle habits can to some degree alter your risk. For example, regular exercise and a healthy low-fat diet rich in fruits and vegetables are related to a lower risk of Alzheimer's disease.

### **Hearing loss**

Studies have found that people who have hearing loss are at risk of dementia. The worse the hearing loss, the higher the risk. However, wearing hearing aids protects against getting dementia.

### **Vision loss that is not treated**

Newer research suggests vision loss that isn't treated is a risk factor for cognitive impairment and dementia. The link may be due to a disease such as diabetes that can increase the risk of both vision loss and dementia. But some research suggests vision loss itself may increase the risk of dementia.

### **Lifelong learning and social engagement**

Studies have found that being social and doing activities that stimulate the mind throughout life can lower the risk of Alzheimer's disease. Low education levels — less than a high school education — appear to be a risk factor for Alzheimer's disease.

## **Symptoms**

Memory loss is the key symptom of Alzheimer's disease. Early in the disease, people may have trouble remembering recent events or conversations. Over time, memory gets worse and other symptoms occur.

At first, someone with the disease may be aware of having trouble remembering things and thinking clearly. As signs and symptoms get worse, a family member or friend may be more likely to notice the issues.

Brain changes from Alzheimer's disease lead to the following symptoms that get worse over time.

### **Memory**

Everyone has trouble with memory at times, but the memory loss related to Alzheimer's disease is lasting. Over time, memory loss affects the ability to function at work and at home.

People with Alzheimer's disease may:

* Repeat statements and questions over and over.
* Forget conversations, appointments or events.
* Misplace items, often putting them in places that don't make sense.
* Get lost in places they used to know well.
* Forget the names of family members and everyday objects.
* Have trouble finding the right words, expressing thoughts or having conversations.

### **Thinking and reasoning**

Alzheimer's disease causes trouble concentrating and thinking, especially about abstract concepts such as numbers. Doing more than one task at once is especially hard. It may be challenging to manage finances, balance checkbooks and pay bills on time. Eventually people with Alzheimer's disease may not recognize numbers.

### **Making judgments and decisions**

Alzheimer's disease makes it hard to make sensible decisions and judgments. People with Alzheimer's disease may make poor choices in social settings or wear clothes for the wrong type of weather. Everyday problems may be hard to solve. Someone with Alzheimer's disease may not know how to handle food burning on the stove or how to make decisions when driving.

### **Planning and performing familiar tasks**

Routine activities that involve completing steps in a certain order also can be hard for people with Alzheimer's disease. They may have trouble planning and cooking a meal or playing a favorite game. As Alzheimer's disease becomes advanced, people forget how to do basic tasks such as dressing and bathing.

### **Changes in personality and behavior**

Brain changes that occur in Alzheimer's disease can affect moods and behaviors. Symptoms may include:

* Depression.
* Loss of interest in activities.
* Social withdrawal.
* Mood swings.
* Not trusting others.
* Anger or aggression.
* Changes in sleeping habits.
* Wandering.
* Loss of inhibitions.
* Delusions, such as believing something has been stolen when it hasn't.

### **Preserved skills**

Despite major changes to memory and skills, people with Alzheimer's disease are able to keep some skills even as symptoms get worse. These are known as preserved skills. They may include reading or listening to books, telling stories, sharing memories, singing, listening to music, dancing, drawing, or doing crafts.

Preserved skills may last longer because they're managed by parts of the brain affected in later stages of the disease.

**What are the early signs of Alzheimer’s?**

In early stages of Alzheimer’s, people often experience memory loss, such as forgetting conversations, events, and the names of familiar people and places. As the disease progresses, the symptoms can include:

* trouble with familiar tasks, such as using a microwave
* difficulties with problem-solving
* trouble with speech or writing

**DIAGNOSIS METHODS**

An important part of diagnosing Alzheimer's disease includes being able to explain your symptoms. It may help to get input from a close family member or friend about your symptoms and their impact on your daily life. Tests of memory and thinking skills also help diagnose Alzheimer's disease.

Blood and imaging tests can rule out other potential causes of your symptoms. They also can check for proteins in the brain that are linked to Alzheimer's disease. The tests may help your healthcare team better identify the disease causing dementia symptoms.

In the past, Alzheimer's disease was diagnosed for certain only after death when plaques and tangles were found while looking at the brain with a microscope. Today, healthcare professionals and researchers are able to diagnose Alzheimer's disease during life with more certainty.

They do this by using a combination of tests that may include tests for biomarkers. Biomarkers can detect if plaques and tangles are present in the brain. Biomarker tests include specific types of positron emission tomography, also known as PET, scans of the brain. Amyloid and tau proteins also can be measured in the fluid part of the blood or in the fluid that surrounds the brain and spinal cord, known as cerebrospinal fluid. Recently, blood biomarker tests have become accurate enough to tell if someone is likely to have amyloid in the brain.

Biomarker tests were mainly used in clinical trials until recently. But healthcare professionals have started using them along with other tests to help diagnose Alzheimer's disease. Biomarker tests also can let healthcare professionals know if the disease is in the early or later stages.

### **Tests**

Diagnosing Alzheimer's disease would likely include the following tests:

### **Physical and neurological exam**

A healthcare professional gives you a physical and neurological exam. This may include testing:

* Reflexes.
* Muscle tone and strength.
* Ability to get up from a chair and walk across the room.
* Sense of sight and hearing.
* Coordination.
* Balance.

### **Lab tests**

Blood tests may help rule out other potential causes of memory loss and confusion, such as a thyroid disorder or vitamin levels that are too low.

Newer blood tests can measure levels of beta-amyloid protein and tau protein. But these tests aren't available everywhere and may not be covered by insurance.

### **Mental status and neuropsychological testing**

Your healthcare professional may give you a brief mental status test to check your memory and other thinking skills. Longer forms of this type of test may provide more details about mental function that can be compared with people of a similar age and education level. These tests can help establish a diagnosis and serve as a starting point to track symptoms in the future.

BRAIN

mages of the brain look for visible changes related to conditions other than Alzheimer's disease that may cause similar symptoms, such as strokes, injury or tumors. Newer imaging tests can help detect specific brain changes caused by Alzheimer's disease, such as amyloid plaques and neurofibrillary tangles. These newer tests are mainly used in major medical centers or in clinical trials.

Imaging of brain structures include:

* **Magnetic resonance imaging (MRI).** MRI uses radio waves and a strong magnetic field to produce detailed images of the brain. They may show shrinkage of some brain regions linked to Alzheimer's disease. MRI scans also can rule out other conditions that may be causing symptoms. An MRI is generally preferred to a CT scan to evaluate dementia. MRIs also are done before starting certain Alzheimer's medicines and throughout treatment to monitor for potential side effects.
* **Computerized tomography (CT).** A CT scan, a specialized X-ray technology, produces cross-sectional images of your brain. It's usually used to rule out tumors, strokes and head injuries.

A PET scan test can capture images of the disease process. During a PET scan, a low-level radioactive tracer is injected into the blood to reveal a particular feature in the brain. PET imaging may include:

* **Fluorodeoxyglucose, also called FDG, PET imaging** scans show areas of the brain where nutrients aren't properly being used for energy, known as metabolism. Finding patterns in the areas of low metabolism can help distinguish between Alzheimer's disease and other types of dementia.
* **Amyloid PET imaging** can measure amyloid plaques in the brain. This test is mainly used in research but may be used if a person has unusual or very early onset of dementia symptoms.
* **Tau PET imaging** measures neurofibrillary tangles in the brain.

Sometimes other tests may be used to measure amyloid and tau in the cerebrospinal fluid. This may be done if symptoms are quickly getting worse or if dementia is affecting someone at a younger age.

### **Future of diagnostic tests**

Research has established that biomarker tests can measure biological signs of disease in the brain. The tests can be used with other tools to help diagnose Alzheimer's disease after symptoms begin. Although these tests can look for signs of Alzheimer's before symptoms begin, the tests aren't being used in people without symptoms. The availability of biomarker tests may vary widely.

Genetic tests aren't recommended for most people who might have Alzheimer's disease. But people with a family history of early-onset Alzheimer's disease may consider being tested. Meet with a genetic counselor to talk about the risks and benefits before getting a genetic test.

**TREATMENT OPTIONS**

*Treatments for Alzheimer's disease include medicines that can help with symptoms and newer medicines that can help slow decline in thinking and functioning. These newer medicines are approved for people with early Alzheimer's disease.*

### ***Medicines***

*Alzheimer's medicines can help with memory symptoms and other cognitive changes. Two types of medicines used to treat symptoms include:*

* ***Cholinesterase inhibitors.*** *These medicines work by boosting levels of cell-to-cell communication. The medicines preserve a chemical messenger that is depleted in the brain by Alzheimer's disease. These are usually the first medicines tried, and most people see modest improvements in symptoms.  
  Cholinesterase inhibitors may improve symptoms related to behavior, such as agitation or depression. The medicines are taken by mouth or through a patch on the skin. Commonly prescribed cholinesterase inhibitors include donepezil (Aricept, Adlarity), galantamine and rivastigmine transdermal patch (Exelon).  
  The main side effects of these medicines include diarrhea, nausea, loss of appetite and trouble with sleep. In people with certain heart conditions, serious side effects may include an irregular heartbeat.*
* ***Memantine (Namenda).*** *This medicine works in another brain cell communication network and slows the progression of symptoms with moderate to severe Alzheimer's disease. It's sometimes used in combination with a cholinesterase inhibitor. Relatively rare side effects include dizziness and confusion.*

*Other medicines have been approved by the Food and Drug Administration, also called FDA, to slow declines in thinking and functioning caused by Alzheimer's disease. The medicines prevent amyloid plaques in the brain from clumping. They're prescribed for people with mild Alzheimer's disease and mild cognitive impairment due to Alzheimer's disease.*

*These medicines include:*

* ***Lecanemab-irmb (Leqembi).*** *This medicine is given as an IV infusion every two weeks. Side effects may include infusion-related reactions such as fever, flu-like symptoms, nausea, vomiting, dizziness, changes in heart rate and trouble breathing.*
* ***Donanemab-azbt (Kisunla).*** *This medicine is given as an IV infusion every four weeks. Side effects may include flu-like symptoms, nausea, vomiting, headache, trouble breathing and changes in blood pressure.*

*People taking lecanemab or donanemab may have swelling in the brain or may get small bleeds in the brain. Rarely, brain swelling can be serious enough to cause seizures and other symptoms. Also in rare instances, bleeding in the brain can cause death. The FDA recommends getting a brain MRI before starting treatment and regularly during treatment to monitor for symptoms of brain swelling or bleeding.*

*People who carry a certain form of a gene known as APOE e4 appear to have a higher risk of these serious side effects. The FDA recommends testing for this gene before starting treatment.*

*If you take a blood thinner or have other risk factors for brain bleeding, talk to your healthcare professional before taking lecanemab or donanemab. Blood-thinning medicines may increase the risk of bleeds in the brain.*

*More research is being done on the potential risks of taking lecanemab and donanemab. Other research is looking at how effective the medicines may be for people at risk of Alzheimer's disease, including people who have a first-degree relative, such as a parent or sibling, with the disease.*

*Sometimes other medicines such as antidepressants may be prescribed to help manage the behavioral symptoms linked to Alzheimer's disease.*

### ***Creating a safe and supportive environment***

*An important part of any treatment plan is to adapt to the needs of a person with Alzheimer's disease. Create routine habits and cut down on tasks that require memory. These steps can make life much easier.*

*These are ways to support a person's sense of well-being and ability to function:*

* *Keep keys, wallets, mobile phones and other valuables in the same place at home so they don't become lost.*
* *Keep medicines in a secure location. Use a daily checklist to keep track of doses.*
* *Arrange for finances to be on automatic payment and automatic deposit.*
* *Have the person with Alzheimer's carry a mobile phone with location tracking. Program important phone numbers into the phone.*
* *Install alarm sensors on doors and windows.*
* *Make sure regular appointments are on the same day at the same time as much as possible.*
* *Use a calendar or whiteboard to track daily schedules. Build the habit of checking off completed items.*
* *Remove furniture, clutter and throw rugs that aren't needed.*
* *Install sturdy handrails on stairs and in bathrooms.*
* *Ensure that shoes and slippers are comfortable and provide good traction.*
* *Reduce the number of mirrors. People with Alzheimer's may find images in mirrors confusing or scary.*
* *Make sure that the person with Alzheimer's carries ID or wears a medical alert bracelet.*
* *Keep photos and other objects with meaning around the house.*

*For Patients, Continue Routine Eye Care*

Though studies are encouraging, there isn’t enough information on the eye-brain connection yet to change anything for patients. If you're worried about memory loss or dementia, talk to your family doctor or see a neurologist. If you have concerns about your vision or eyes, see an ophthalmologist.

**PREVENTION TIPS**

Alzheimer's disease can't be prevented. But making lifestyle changes can lower your risk of getting the disease.

Research suggests that taking steps that lower your risk of cardiovascular disease may reduce the risk of dementia, as well. To follow heart-healthy lifestyle choices that may lower the risk of dementia:

* Exercise regularly.
* Eat a diet of fresh produce, healthy oils and foods low in saturated fat, such as a Mediterranean diet.
* Work with your healthcare professional to manage high blood pressure, diabetes and high cholesterol. Pay particular attention to your levels of low-density lipoprotein, known as LDL, cholesterol. High levels of LDL cholesterol in people younger than 65 raises the risk of dementia. But taking medicines to lower LDL cholesterol doesn't raise the risk.
* If you smoke, ask your healthcare professional for help to quit.

One large, long-term study done in Finland found that making lifestyle changes helped reduce cognitive decline among people who were at risk of dementia. Those in the study were given individual and group sessions that focused on diet, exercise and social activities.

Several studies have found that following a Mediterranean diet leads to better cognitive function and a slower cognitive decline with age. A Mediterranean diet focuses on plant-based foods such as fruits, vegetables, grains, fish, poultry, nuts and olive oil. The diet includes less foods that are high in saturated fats and trans fats, such as butter, margarine, cheese, red meat, fried food and pastries.

It's also important to treat vision loss and hearing loss. Studies have found that vision loss that isn't treated is a risk factor for cognitive impairment and dementia. Studies also have found that people who have hearing loss are at higher risk of dementia. But wearing hearing aids made people less likely to get dementia.

Other studies have shown that staying engaged mentally and socially is linked to preserved thinking skills later in life and a lower risk of Alzheimer's disease. This includes going to social events, reading, dancing, playing board games, creating art, playing an instrument and other activities.

**WHEN TO SEE A DOCTOR / RED FLAG**

You should speak with a doctor if you’re having problems with memory loss that are out of the ordinary for you and experiencing difficulties with thinking. It’s also a good idea to talk with a doctor if relatives or friends who spend a lot of time around you have noticed a decrease in your memory.

**COMPLICATION**

Alzheimer's disease can lead to a variety of complications. Symptoms such as memory loss, language loss, impaired judgment and other brain changes can make it harder to manage other health conditions. A person with Alzheimer's disease may not be able to:

* Tell someone about being in pain.
* Explain symptoms of another illness.
* Follow a treatment plan.
* Explain medicine side effects.

As Alzheimer's disease moves into its last stages, brain changes begin to affect physical functions. The changes can affect the ability to swallow, balance, and manage stool and bladder movements. These effects can lead to other health issues such as:

* Inhaling food or liquid into the lungs.
* Flu, pneumonia and other infections.
* Falls.
* Fractures.
* Bedsores.
* Poor nutrition or dehydration.
* Constipation or diarrhea.

**Alternative medicine**

Herbal remedies, vitamins and other supplements are widely promoted for cognitive health or to prevent or delay Alzheimer's. But clinical trials have produced mixed results. There's little evidence to support them as effective treatments.

Some of the treatments that have been studied include:

* **Vitamin E.** Although vitamin E doesn't prevent Alzheimer's, taking 2,000 international units daily may help delay symptoms from getting worse in people who already have mild to moderate disease. But study results have been mixed, with only some showing modest benefits. Further research into the safety of people with dementia taking 2,000 international units daily of vitamin E are needed before it can be routinely recommended.  
  Supplements promoted for cognitive health can interact with medicines you're taking for Alzheimer's disease or other health conditions. Work closely with your healthcare team to create a safe treatment plan. Tell your healthcare team about your prescriptions and any medicines or supplements taken without a prescription.
* **Omega-3 fatty acids.** Omega-3 fatty acids in fish or from supplements may lower the risk of dementia. But clinical trials have shown no benefit for treating Alzheimer's disease symptoms.
* **Curcumin.** This herb comes from turmeric and has anti-inflammatory and antioxidant properties that might affect chemical processes in the brain. So far, clinical trials have found no benefit for treating Alzheimer's disease.
* **Ginkgo.** Ginkgo is a plant extract. A large study funded by the National Institutes of Health found no effect in preventing or delaying Alzheimer's disease.
* **Melatonin.** This supplement helps with sleep. It's being studied to see if it can help people with dementia manage sleep symptoms. But some research has found that melatonin may worsen mood in some people with dementia. More research is needed.

**Lifestyle and home remedies**

Healthy lifestyle choices promote good overall health. They also may play a role in maintaining brain health.

### **Exercise**

Regular exercise is an important part of a treatment plan. Activities such as a daily walk can help improve mood and maintain the health of joints, muscles and the heart. Exercise also promotes restful sleep and prevents constipation. It's beneficial for care partners, too.

People with Alzheimer's who have trouble walking may still be able to use a stationary bike, stretch with elastic bands or do chair exercises. You might find exercise programs for older adults at community centers or on TV, the internet or DVDs.

### **Nutrition**

People with Alzheimer's may forget to eat, lose interest in meals or may not eat healthy foods. They may also forget to drink enough, leading to dehydration and constipation.

Offer the following:

* **Healthy options.** Buy favorite healthy food options that are easy to eat.
* **Water and other healthy beverages.** Encourage drinking several glasses of liquids every day. Don't offer beverages with caffeine, which can increase restlessness, affect sleep and trigger a need to urinate often.
* **High-calorie, healthy shakes and smoothies.** Serve milkshakes with protein powders or make smoothies. This is helpful when eating becomes very hard.

### **Social engagement and activities**

Social activities can support preserved skills and abilities. They also help with overall well-being. Do things that are meaningful and enjoyable. Someone with dementia might:

* Listen to music or dance.
* Read or listen to books.
* Garden or do crafts.
* Go to social events at senior or memory care centers.
* Do activities with children.

**Differential diagnosis (how it’s distinguished from other illnesses)**

## Normal Aging

* Mild forgetfulness and slower cognitive processing can occur with normal aging.
* Does not significantly impair daily functioning or cause progressive decline as in AD.

## 2. Other Dementias

* Vascular Dementia
  + Cognitive decline associated with cerebrovascular disease (strokes, small vessel disease).
  + Often has a stepwise progression and focal neurological signs.
  + Imaging shows infarcts or white matter changes.
* Dementia with Lewy Bodies (DLB)
  + Fluctuating cognition, visual hallucinations, parkinsonism.
  + Early attention and visuospatial deficits.
* Frontotemporal Dementia (FTD)
  + Early changes in personality, behavior, and language rather than memory.
  + Younger age of onset than AD.
* Parkinson’s Disease Dementia
  + Cognitive decline occurring after established Parkinson’s disease.
* Creutzfeldt-Jakob Disease and Other Prion Diseases
  + Rapidly progressive dementia with motor signs and myoclonus.

## 3. Mild Cognitive Impairment (MCI)

* Memory impairment without significant impact on daily function.
* Amnestic MCI often precedes AD but does not always progress to dementia.

## 4. Delirium and Acute Confusional States

* Sudden onset cognitive impairment often reversible.
* Associated with infection, metabolic disturbances, medications.

## 5. Depression (“Pseudodementia”)

* Cognitive symptoms due to depression can mimic dementia.
* History of mood symptoms and response to antidepressants help differentiate.

## 6. Metabolic and Endocrine Disorders

* Hypothyroidism, vitamin B12 deficiency, and other metabolic causes can cause reversible cognitive impairment.

## 7. Infections and Inflammatory Conditions

* Neurosyphilis, HIV-associated dementia, autoimmune encephalitis.

## 8. Drug-Induced Cognitive Impairment

* Benzodiazepines, anticholinergics, and other medications.

Clinical and Diagnostic Tools to Differentiate AD

* History and Neuropsychological Testing
  + AD typically presents with insidious onset and progressive episodic memory loss.
  + Impairment in at least two cognitive domains affecting daily function is required.
* Neuroimaging
  + MRI or CT: hippocampal and cortical atrophy in AD; vascular lesions in vascular dementia.
  + PET/SPECT: hypometabolism or hypoperfusion in temporoparietal regions in AD.
* Laboratory Tests
  + To exclude reversible causes (thyroid function, B12, syphilis serology, HIV).
* Biomarkers (mainly research use)
  + CSF amyloid beta and tau proteins.
  + Amyloid and tau PET imaging.

**RECENT GUIDELINES OR UPDATES**

**New Research Points In Promising Directions**

* Alzheimer’s Association Clinical Practice Guidelines (2025)  
  The Alzheimer’s Association is publishing its first comprehensive clinical practice guideline in 2025 focused on the use of blood biomarkers for AD diagnosis in patients with cognitive impairment seen in specialty care settings. This guideline will:
  + Provide recommendations on using blood biomarker tests as confirmatory or triaging tools in the diagnostic workup.
  + Support evidence-based clinical decision making and shared decision making with patients.
  + Be regularly updated to incorporate new evidence and expand to additional patient populations and care settings.
* Structured Diagnostic Evaluation  
  New evidence-based guidelines empower clinicians—including primary care providers—to implement structured, patient-centered evaluations for cognitive impairment potentially due to AD or related dementias. These include:
  + Use of validated cognitive, mood, behavioral, and functional assessment tools.
  + Integration of clinical findings with biomarker data (CSF, PET imaging, blood tests) when available.
  + Guidance on differentiating AD from other dementias such as Lewy body dementia and frontotemporal dementia.
* Biomarker and Imaging Integration  
  Guidelines increasingly recommend incorporating biomarkers (amyloid beta, tau) and neuroimaging (MRI, PET) to improve diagnostic accuracy, especially in early or atypical cases.
* Risk-Based Approach in Asymptomatic Individuals  
  Updated international recommendations suggest classifying cognitively normal individuals with positive AD biomarkers as “at risk” rather than diagnosing them with AD. This avoids unnecessary labeling and focuses on monitoring and preventive strategies.
* Prevention and Staging Guidelines  
  Additional forthcoming guidelines will address:
  + Use of cognitive assessment tools in primary care for early detection of mild cognitive impairment (MCI) and dementia.
  + Staging of AD to guide prognosis and management.
  + Prevention strategies targeting modifiable risk factors.
* Clinical Practice Guideline Development Process  
  The Alzheimer’s Association uses rigorous, transparent methods (GRADE framework) involving multidisciplinary experts and patient representatives, with public comment opportunities, to ensure trustworthy, actionable recommendations.

**Epidemiology data**

* Prevalence in the United States:  
  Approximately 7.2 million Americans aged 65 and older are living with Alzheimer’s disease in 2025, representing about 1 in 9 people in this age group. Nearly three-quarters (74%) of those affected are aged 75 or older. Women constitute almost two-thirds of cases, with a lifetime risk at age 45 of about 1 in 5 for women and 1 in 10 for men.
* Projected Growth:  
  The number of Americans with Alzheimer’s is expected to nearly double by 2050, reaching about 13 to 13.8 million people aged 65 and older unless effective prevention or cures are developed. Globally, over 55 million people were estimated to have Alzheimer’s or other dementias in 2018, with projections exceeding 152 million by 2050.
* Incidence and Age Distribution:  
  Alzheimer’s primarily affects older adults, with incidence increasing sharply with age. After age 80, about 1 in 6 people develop Alzheimer’s dementia. While less common, younger-onset Alzheimer’s (before age 65) affects approximately 200,000 Americans.
* Racial and Ethnic Disparities:  
  Older Black Americans are about twice as likely, and older Hispanic Americans about 1.5 times as likely, to have Alzheimer’s or other dementias compared to older Whites. Despite higher prevalence, Black and Hispanic individuals are less likely to be diagnosed.
* Economic and Caregiving Burden:  
  In 2025, health and long-term care costs for people with dementia in the U.S. are projected to reach $384 billion, rising to nearly $1 trillion by 2050. Unpaid caregivers provide nearly 19 billion hours of care annually, valued at over $413 billion.
* Mortality:  
  Alzheimer’s disease is among the top 10 leading causes of death in the U.S. and is the only leading cause of death that continues to increase. Deaths attributed to Alzheimer’s increased by 145% between 2000 and 2019.
* Global Perspective:  
  About 60% of people with dementia live in low- and middle-income countries, a proportion expected to rise to 71% by 2050. Worldwide, more than 10 million new dementia cases are diagnosed annually, equating to one new case every 3.2 seconds

REFERENCES: <https://www.healthline.com/health/alzheimers-disease-risk-factors#faq>

<https://pmc.ncbi.nlm.nih.gov/articles/PMC5501038/>

**AMBLYOPIA (LAZY EYE)**

**DEFINITION / DESCRIPTION**

Amblyopia (Lazy Eye) is a condition where one or both eyes does not develop normal vision during childhood, often due to misalignment of the eyes, a difference in focus between the eyes, or improper brain-eye coordination, or maybe the child’s brain learnt to ignore one of the eyes; it is often caused by strabismus or refractive differences between eyes. Reduced vision in one eye due to. One of the most common causes of vision impairment in children .

It is sometimes called “lazy eye.” Amblyopia is a common problem in babies and young children, but vision changes from amblyopia can last a lifetime.

A child’s vision develops in the first few years of life. It is important to diagnose and treat amblyopia as early as possible. Otherwise, a child with amblyopia will not develop normal, healthy vision.

Lazy eye (amblyopia) is reduced vision in one eye caused by abnormal visual development early in life. The weaker — or lazy — eye often wanders inward or outward.

Amblyopia generally develops from birth up to age 7 years. It is the leading cause of decreased vision among children. Rarely, lazy eye affects both eyes.

Early diagnosis and treatment can help prevent long-term problems with your child's vision. The eye with poorer vision can usually be corrected with glasses or contact lenses, or patching therapy.

**CAUSES**

Lazy eyes develop because of abnormal visual experience early in life that changes the nerve pathways between a thin layer of tissue (retina) at the back of the eye and the brain. The weaker eye receives fewer visual signals. Eventually, the eyes' ability to work together decreases, and the brain suppresses or ignores input from the weaker eye.

Anything that blurs a child's vision or causes the eyes to cross or turn out can result in a lazy eye. Common causes of the condition include:

* **Muscle imbalance (strabismus amblyopia).** The most common cause of lazy eye is an imbalance in the muscles that position the eyes. This imbalance can cause the eyes to cross in or turn out, and prevents them from working together.
* **Difference in sharpness of vision between the eyes (refractive amblyopia).** A significant difference between the prescriptions in each eye — often due to farsightedness but sometimes to nearsightedness or an uneven surface curve of the eye (astigmatism) — can result in lazy eye.

Glasses or contact lenses are typically used to correct these refractive problems. In some children, a lazy eye is caused by a combination of strabismus and refractive problems.

* **Deprivation.** A problem with one eye — such as a cloudy area in the lens (cataract) — can prohibit clear vision in that eye. Deprivation amblyopia in infancy requires urgent treatment to prevent permanent vision loss. It's often the most severe type of amblyopia.

**RISK FACTORS**

Several risk factors contribute to the development of amblyopia/lazy eye include:

* Premature birth
* Small size at birth
* Family history of lazy eye
* Developmental disabilities
* Ptosis
* Strabismus
* Childhood glaucoma
* Early-onset cataracts
* Uveitis
* Systemic conditions with ocular manifestation
* Children born small for gestational age
* Low birth weight (less than 1500 g)
* Premature birth (gestational age less than 30 weeks)
* Developmental delay
* First-degree relatives with the condition
* Maternal use of smoking
* Alcohol or drug use during pregnancy

**SIGNS / SYMPTOMS**

Signs and symptoms of lazy eye include:

* An eye that wanders inward or outward
* Eyes that appear to not work together
* Poor depth perception
* Squinting or shutting an eye
* Head tilting
* Abnormal results of vision screening tests

Sometimes a lazy eye is not evident without an eye exam.

**DIAGNOSIS METHODS**

Ophthalmologists diagnose amblyopia by checking to see if vision differs between the two eyes. To check a baby's or young child's vision, the ophthalmologist may cover one of the child's eyes and watch how well they can follow a moving object. The doctor may also watch how the child reacts when one eye is covered. If one eye has amblyopia and the other is covered, the child may try to look above or below the patch, pull it off or cry.

The ophthalmologist will do a complete medical eye exam, looking for other eye problems that could be affecting vision.

**When Should a Child's Vision Be Tested?**

All children should have their vision checked by their pediatrician or family physician during their routine doctor's appointments. Many children will also have their vision checked at school.   
 If a child's doctor, school, or family is concerned about their vision, the child should see an ophthalmologist for further tests.

**TREATMENT OPTIONS**

It's important to start treatment for lazy eye as soon as possible in childhood, when the complicated connections between the eye and the brain are forming. The best results occur when treatment starts before age 7, although half of children between the ages of 7 and 17 respond to treatment.

Treatment options depend on the cause of the lazy eye and on how much the condition is affecting your child's vision. Your doctor might recommend:

* **Corrective eyewear.** Glasses or contact lenses can correct problems such as nearsightedness, farsightedness or astigmatism that result in lazy eye.
* **Eye patches.** To stimulate the weaker eye, your child wears an eye patch over the eye with better vision for two to six or more hours a day. In rare cases, wearing an eye patch too long can cause amblyopia to develop in the patched eye. However it's usually reversible.
* **Bangerter filter.** This special filter is placed on the eyeglass lens of the stronger eye. The filter blurs the stronger eye and, like an eye patch, works to stimulate the weaker eye.
* **Eyedrops.** An eyedrop of a medication called atropine (Isopto Atropine) can temporarily blur the vision in the stronger eye. Usually prescribed for use on weekends or daily, use of the drops encourages your child to use the weaker eye, and offers an alternative to a patch. Side effects include sensitivity to light and eye irritation.
* **Surgery.** Your child might need surgery if he or she has droopy eyelids or cataracts that cause deprivation amblyopia. If your child's eyes continue to cross or wander apart with the appropriate glasses, your doctor might recommend surgical repair to straighten the eyes, in addition to other lazy eye treatments.

Activity-based treatments — such as drawing, doing puzzles or playing computer games — are available. The effectiveness of adding these activities to other therapies hasn't been proved. Research into new treatments is ongoing.

For most children with lazy eyes, proper treatment improves vision within weeks to months. Treatment might last from six months to two years.

It's important for your child to be monitored for recurrence of lazy eye — which can happen in up to 25 percent of children with the condition. If lazy eye recurs, treatment will need to start again.

**How to Choose and Use an Eye Patch**

An eye patch should be comfortable yet remain firmly in place. It should also not allow the child to peek around its edges. Most drug stores have a variety of sizes and types of eye patches. Decorated fun patches are available online. Do not use the black eye patches with elastic bands or ties (such as pirate-type patches). These are too easy for a child to remove or peek around. To wear the patch, simply attach it to the skin around your child’s eye.

If your child wears glasses, there are patches designed to attach to the lens. These may be good for children who are used to wearing a patch, but they are not as good for a child new to treatment. This is because the patch can slip or the child may learn to peek around it. If your child wears glasses and is not used to patching, it is best to attach the patch directly around the stronger eye underneath the glasses.

**Keep Your Child from Taking Off the Eye Patch**

Children do not like to have their stronger eyes patched or blurred. However, you need to help your child do what is best for them. Otherwise, treatment will not work.

Try distracting the child or having them do something that keeps their attention. Or reward the child with a treat for wearing the patch.

It can take a while for your child to get used to wearing a patch. Over time, this should get easier for them and you. Remember that strengthening the weaker eye is the only way to develop healthy, normal vision.

If your child still takes off the patch, as a last resort, you might cover his or her hands with gloves, mittens, or socks.

**Teach Your Child About the Eye Patch**

Pre-school or school-age children might not want to wear an eye patch or use blurring eye drops. To help, parents should explain how important these treatments are to be able to see well. And reassure them that lots of children wear eye patches for the same reason.

Consider having a very young child practice putting an eye patch on a doll. Or let the child decorate his or her patch with crayons or markers.

Explain the amblyopia treatment to the child’s teacher. Ask the teacher to compliment the child on being so good about wearing the patch. Children thrive on positive feedback from their teachers.

Things to consider with patching treatment:

* In very rare instances it is possible to overuse the patch or blurring eye drops. This can affect vision in the stronger eye. Be sure to keep the child’s appointments with the ophthalmologist so that vision in both eyes can be closely monitored.
* The skin near your child’s eye patch can get irritated. To help, try a different size or type of patch, and angle the patch differently each day.
* Your child may initially be clumsy when wearing a patch. Try to keep an eye on your child when they are climbing stairs or being active.

**Treating Amblyopia Using New Technology**

A new treatment for amblyopia uses a virtual reality (VR) headset to help improve vision in children aged 4 to 7. A child watches videos wearing the headset, which helps them use their weaker eye. To learn more, ask your child’s ophthalmologist.

**Treating Amblyopia for Better Lifelong Vision**

When a child has amblyopia, it is important to make vision stronger in the weak eye. Even if eye problems causing amblyopia are corrected with glasses or surgery, the amblyopia itself must be treated. If not, the child may have lifelong vision problems.

**PREVENTION TIPS**

You can’t prevent amblyopia or the other vision issues that cause it. The best thing you can do for your child’s eyes and vision is to have them checked regularly.

**OUTLOOK / PROGNOSIS**

Amblyopia is very treatable if it’s diagnosed early. Children with amblyopia who start treatment early in life are much more likely to have improved vision and fewer long-term effects.

Your eye care specialist will suggest treatments that improve your child’s sight as much as possible. They might need vision correction lenses like glasses or contact lenses their whole life.

**Can my child grow out of amblyopia?**

No, amblyopia doesn’t go away on its own and children can’t grow out of it. If it’s not treated, amblyopia can cause permanent vision issues, including blindness in the affected eye.

**Living With**

Having your child’s eyes and vision checked regularly can help an eye care specialist identify problems right away. A pediatrician should check your child’s eyes at every well-child visit until they’re old enough to start school, and then every one to two years.

**POSSIBLE COMPLICATIONS**

Untreated, lazy eye can cause permanent vision loss

**WHEN TO SEE A DOCTOR**

See your child's doctor if you notice his or her eye wandering after the first few weeks of life. A vision check is especially important if there's a family history of crossed eyes, childhood cataracts or other eye conditions.

For all children, a complete eye exam is recommended between ages 3 and 5.

**DIFFERENTIAL DIAGNOSIS**

* A-Pattern Esotropia and Exotropia
* Accommodative Esotropia
* Acquired Esotropia
* Acquired Exotropia
* Congenital Exotropia
* Congenital Ptosis (Drooping Eyelid)
* Esotropia with High AC/A Ratio
* Infantile Esotropia
* Monofixation Syndrome
* V-Pattern Esotropia and Exotropia

**EPIDEMIOLOGY**

Amblyopia presents a significant global health concern owing to its prevalence and the potential for permanent visual impairment if it is not promptly diagnosed and treated. Historically, various forms of amblyopia have been reported to affect up to 3% of the population, with a 1.2% lifetime risk of vision loss attributed to this condition. More recent data suggest that the global prevalence of amblyopia falls within the range of 1.1% to 1.8%.

Population-based studies have reported varying prevalence rates, with estimates ranging from 0.7% to 2.6% among children aged 30 to 70 months and 1.0% to 5.5% in older children. The reported prevalence of amblyopia can fluctuate based on factors such as age, ethnicity, race, the specific definition used for amblyopia, the methodology of the study, and other contributing variables.

A comprehensive meta-analysis from global prevalence data encompassing more than 1.8 million patients across 60 studies revealed a pooled prevalence rate of 1.44%, with a range of 1.17% to 1.78%. The prevalence rates varied across different continents, with Europe, North America, Asia, and Africa reporting rates of 2.90%, 2.41%, 1.09%, and 0.72%, respectively. The study estimated that in 2019, approximately 99.2 million people worldwide were affected by amblyopia, with a projected increase to 175 million by the year 2030 and 220 million by 2040.

Amblyopia typically manifests as a unilateral condition. However, there are instances where patients can develop the bilateral form if both eyes experience visual alterations during early development. Anisometropia is the most common cause of amblyopia, followed by mixed anisometropia and strabismus, strabismus, and visual deprivation. Studies have indicated that the relative prevalence rates, categorized by the type of amblyopia, are approximately 50% due to anisometropia, 19% linked to strabismus, 27% arising from a combination of both factors, and 4% attributed to deprivation-related causes.

The likelihood of developing amblyopia increases significantly with specific factors. When the difference in refraction error between the eyes falls within the range of 1D to 2D of spherical equivalence, the odds of developing amblyopia are 4.5 times higher. This risk can escalate to a 40-fold increase when the difference in refraction error exceeds 2D.

Regarding strabismus, amblyopia risk has been reported to be between 3 and 18 times greater compared to individuals without strabismus. Mixed and strabismic amblyopia is typically diagnosed at an earlier age, with an average age of 7.4 years, compared to anisometropic amblyopia, which is usually diagnosed at an average age of 12.7. The prevalence of amblyopia appears to be similar between right and left eyes, and there is no observed gender preference.

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**ANISOCORIA**

*ALTERNATIVE NAMES:* Anisocoria is also referred to as “unequal pupil size” or “pupil asymmetry”

**DEFINITION / DESCRIPTION**

Anisocoria is when your eye’s pupils are not the same size. The pupil allows light to enter the eye so that you can see.

Anyone can have pupils that differ in size with no problems. In fact, 1 out of 5 people have pupils that are normally different sizes.

Sometimes, though, having uneven pupil size can be a symptom of a serious eye problem. People who may get anisocoria include those with:

* a nervous system problem
* a history of damage to the eye
* risk of having a stroke
* a viral infection
* Adie’s tonic pupil (when one pupil does not respond to light as well as the other pupil)

Anisocoria is the medical term for one of your pupils being bigger than the other. The pupil is the black center of your eye that changes size to help you see in different amounts of light. It shrinks (contracts) in bright light and expands (dilates) in dim light.

Your pupils automatically adjust throughout the day without you noticing or controlling them. If you have anisocoria, one will be stuck noticeably larger than the other. You’ll probably be able to see the size difference in a mirror or selfie.

This might affect your vision. If one pupil can’t adjust to light like it should, you might have trouble seeing clearly, or be sensitive to light in your affected eye.

Visit an eye care specialist or go to the emergency room if you notice one of your pupils is suddenly larger than the other. Some people develop anisocoria with no long-term complications, but it can also be a sign of a life-threatening medical emergency. If you have other symptoms, like pain or noticeably worse vision, go to your nearest emergency room.

**Types of anisocoria**

Eye care specialists classify anisocoria as either physiological (caused by something malfunctioning inside your body) or pathological (caused by a health condition). Physiological anisocoria is more common.

This distinction isn’t as important as getting your symptoms evaluated right away. No matter what’s causing your pupils to be irregularly sized, you should see an eye care specialist as soon as possible.

Experts estimate that around 15% of people experience anisocoria at some point in their lives. Some babies are born with it (congenital anisocoria), but it usually happens because of a health condition or another issue that affects your eyes.

**CAUSES**

Anisocoria can be caused by lots of health conditions and injuries, and can be a side effect of some medications. It can also happen without a known cause (idiopathic anisocoria). Some of the most common causes include:

* Migraine headaches.
* Surgery side effects.
* Eye drops, scopolamine skin patches and some other medicines.
* Eye injuries and traumas that affect your head, eyes or the arteries that supply blood to them.

Some serious, potentially fatal health conditions can cause anisocoria, including:

* Brain aneurysms.
* Strokes.
* Brain tumors.
* Infections like meningitis.
* Certain types of cancer.

**SIGNS / SYMPTOMS**

One pupil being noticeably bigger (more dilated) than the other is the most obvious anisocoria symptom. You might not experience other symptoms. If that’s the case, you still need to visit an eye care specialist to have the change in your eyes diagnosed.

Go to the emergency room if one pupil is bigger than the other and you experience any of the following:

* Eye pain.
* Blurry vision.
* Double vision (diplopia).
* Light sensitivity (photophobia).
* Sudden vision loss.

Other symptoms can include:

* Fever.
* Headaches.
* Nausea or vomiting.
* Neck pain or stiffness.

**ANISOCORIA DIAGNOSIS METHOD**

To diagnose anisocoria, your ophthalmologist will examine your pupils in both a lighted room and a dark room. This allows them to see how your pupils respond to light. This can help them figure out which pupil is abnormal.

Your ophthalmologist will also check your eyes with a slit-lamp microscope. This instrument lets your eye doctor look at your eye in small, detailed sections. That makes it easier to spot problems.

If you have other symptoms along with different pupil size, your ophthalmologist will do other tests to learn more about your condition.

An eye care specialist or another healthcare provider will diagnose anisocoria with an eye exam. They’ll perform a physical exam to check for other symptoms. They might give you eye drops to make it easier to examine your eyes.

Your provider may use imaging tests to look for what’s causing anisocoria, including:

* MRI (magnetic resonance imaging).
* CT scan (computed tomography scan).
* X-ray.

You might also need blood tests or a lumbar puncture if your provider thinks you have an infection.

**ANISOCORIA TREATMENT OPTIONS**

Finding and treating what’s causing the anisocoria is more important than managing the irregularity in your pupils.

If you have no other symptoms — and your eye care specialist rules out any serious underlying conditions — you probably won’t need any treatment. Your pupils may return to their usual size over time.

If anisocoria is the first sign of a more serious condition, the treatment you’ll need depends on what’s causing it. Talk to your provider about what you’ll need to do next and what to expect.

**PREVENTION TIPS**

Because lots of conditions and injuries can cause anisocoria, there’s not one surefire way to prevent it.

Have your eyes examined regularly and see a healthcare provider every year for a checkup. Regular eye exams and maintaining your overall health are the best ways to catch issues that might cause anisocoria before they damage your eyes and body.

**OUTLOOK / PROGNOSIS**

Everyone’s outlook is different. It depends on what’s causing the anisocoria. Some issues like migraines or reactions to medications will go away on their own. If you experience something more serious like an aneurysm or stroke, your life might be changed permanently.

For most people, anisocoria is a minor part of a larger health issue, and as you treat your underlying condition, your pupils will return to their usual size. Ask your healthcare provider or eye care specialist about your unique outlook.

**Living With**

Visit an eye care specialist as soon as you notice any changes in your eyes or vision.

Go to the emergency room right away if your pupils are different sizes and you have sudden, new pain or vision loss.

**DIFFERENTIAL DIAGNOSIS**

The differential of conditions causing anisocoria may be divided into three sub-categories, depending upon whether the anisocoria increases in dim light, in bright light, or remains equal in both lighting conditions.

## **Differential Diagnoses**

* Basilar Artery Thrombosis
* Brainstem Gliomas
* Cardioembolic Stroke
* Cavernous Sinus Syndromes
* Cerebral Aneurysm
* Cluster Headache
* Diagnosis and Management of Cervical Spondylosis
* Dissection Syndromes
* Migraine Headache
* Migraine Variants
* Migraine in Children
* Multiple Sclerosis
* Neuro-Ophthalmic Examination
* Neuro-Ophthalmic History
* Neuro-ophthalmic Manifestations of Vascular Eye Diseases
* Raeder Paratrigeminal Syndrome

**EPIDEMIOLOGY**

The prevalence of physiologic anisocoria is generally considered to be around 10 to 20%, which does not seem to differ greatly around the world. Physiologic anisocoria does not seem to have a sex predilection nor occurs at a specific age. Physiologic anisocoria is probably the most common cause. The prevalence of other causes of anisocoria is associated with the prevalence of the underlying condition.

### Frequency

United States

Anisocoria is common, although no overall prevalence statistics are available. The incidence and prevalence data for anisocoria vary by the specific pathophysiology and population. The presence of physiologic anisocoria has been estimated at 20% of the normal population, so some degree of pupil difference may be expected in at least 1 in 5 clinic patients.

### Mortality/Morbidity

Mortality and morbidity rates associated with anisocoria depend entirely upon the specific pathophysiology.

Several causes of anisocoria are life threatening, including Horner syndrome due to carotid dissection or third nerve palsy due to an aneurysm or uncal herniation. Anisocoria may also be caused by sight-threatening origins, such as angle closure glaucoma. Yet other causes of anisocoria are completely benign (eg, simple or physiologic anisocoria), although the unnecessary evaluation of these disorders may produce morbidity inadvertently.

REFERENCES

[**Anisocoria (Unequal Pupil Size): Symptoms & Causes**](https://my.clevelandclinic.org/health/diseases/22422-anisocoria)

[**Anisocoria Differential Diagnoses**](https://emedicine.medscape.com/article/1158571-differential?form=fpf)

**ANIRIDIA**

*ALTERNATIVE NAMES:* Aniridia is also known as “absent iris”, or “without iris”, “congenital aniridia”, and “irideremia”

**DEFINITION / DESCRIPTION**

Aniridia is a condition that causes babies to be born without [irises](https://my.clevelandclinic.org/health/body/22502-iris) (the colored part of your eye). Some babies are missing their entire irises. Others only have part of an iris in each eye.

Aniridia always affects both of your child’s eyes (it’s a bilateral condition).

A healthcare provider will probably diagnose aniridia when your child is born. They’ll be able to see the missing iris in your child’s eyes. No matter how much of your baby’s irises are missing, aniridia will affect their vision and can eventually lead to other issues in their eyes later in life.

If your child has aniridia, their pupils will look larger than usual

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**CAUSES OF ANIRIDIA**

Aniridia is a genetic disorder. It happens when a genetic change affects the *PAX6* gene. This gene helps form your baby’s eyes and parts of their brain, spinal cord and pancreas. The genetic change that causes aniridia usually happens between the 12th and 14th week of pregnancy.

**RISK FACTORS**

Aniridia is more common in children whose biological parents have it. If one parent has aniridia, there’s a 50/50 chance their biological child will, too.

So, if you have aniridia, that doesn’t guarantee that your biological children will have it. It just means they’re more likely to.

Aniridia can also happen on its own (sporadically) even if neither biological parent has it. This happens in around 1 in 3 cases.

**SIGNS / SYMPTOMS**

Because your child is missing their irises, their pupils will look larger than usual. Their pupils might be unevenly shaped. They’ll have a hard time adjusting to changes in lighting. This can lead to symptoms like:

* Blindness or partial vision loss in one or both eyes
* Blurry vision
* Eye pain
* Low vision
* Light sensitivity (photophobia)

**DIAGNOSIS METHODS**

Your provider will diagnose aniridia when your baby is born. You should be able to see the missing irises in your baby’s eyes.

If you’re pregnant and concerned about the fetus’s risk of aniridia (or other genetic disorders), talk to your provider about prenatal genetic testing. Your provider will use a blood test to tell how likely it is that the fetus could have a genetic disorder. They might also perform amniocentesis — removing and testing a small amount of your amniotic fluid.

**TREATMENT OPTIONS**

Treating aniridia focuses on maintaining or improving your child’s vision.

Your child will need regular eye exams and visits with an eye care specialist. The sooner your provider diagnoses changes in your child’s eyes, the more likely they’ll be able to prevent symptoms or complications.

Your child might need a few treatments, including:

* Glasses or contact lenses. Just like anyone with vision issues, wearing glasses or contact lenses can improve your child’s vision. Kids with aniridia sometimes wear specialized colored contacts that mimic the shape of an iris to cover their pupils and reduce light sensitivity.
* Medications. If your child develops glaucoma or cornea issues, your provider might prescribe medicated eye drops, artificial tears or other medications.
* Surgery. Children who develop cataracts may need cataract surgery to remove them. Your child might also need glaucoma surgery. They might be able to get implanted artificial irises. This is a relatively new treatment, so not every child is a good candidate for this type of surgery.
* Prevention tips

**OUTLOOK / PROGNOSIS**

You should expect your child to have vision issues. Your child will need regular eye exams throughout their life to monitor their eye health.

More than 8 in 10 children with aniridia have poor or impaired eyesight. Most people with aniridia develop glaucoma, usually when they’re 10 to 20 years old.

But this doesn’t mean every baby born with aniridia will definitely face these issues. Talk to your provider or eye care specialist about what to expect as your child gets older.

**POSSIBLE COMPLICATIONS**

Other parts of your child’s eyes might be underdeveloped, as well, including their optic nerves and retinas.

Children with aniridia are likely to develop other eye issues as they grow up, including:

* Cataracts
* Cloudy corneas
* Glaucoma
* Nystagmus

Children with sporadic aniridia have a higher risk of developing a Wilms tumor (a rare type of kidney cancer). The genetic change that causes sporadic aniridia is much more likely to impact other parts of your child’s body that the *PAX6* gene is responsible for.

**WHEN TO SEE A DOCTOR / RED FLAG**

See a healthcare provider as soon as you notice any changes in your child’s eyes or vision. You might want to ask your provider questions like:

* How often will my child need eye exams?
* Is my child a good candidate for artificial iris surgery?
* What are the chances my child experiences complications?
* Which changes or symptoms should I look out for?

Go to the emergency room if your child has any of the following symptoms:

* A sudden loss of vision
* Severe eye pain
* Seeing new flashes or floaters in their eyes

**DIFFERENTIAL DIAGNOSIS**

* Congenital Cataract
* Congenital Nystagmus
* Ectopia Lentis
* Iris Coloboma
* Juvenile Glaucoma

**EPIDEMIOLOGY**

Aniridia is noted in around 1.8 of 100,000 births. Males and females are equally affected.

* Prevalence:  
  The prevalence of aniridia ranges from approximately 1 in 40,000 to 1 in 100,000 individuals worldwide, with no gender predilection. Specific population-based data include:
  + Sweden: about 1 in 70,000
  + Norway: about 1 in 76,000
  + General estimate: around 1.8 per 100,000 newborns[7](https://www.news-medical.net/health/What-is-Aniridia.aspx).
* Inheritance Patterns:  
  About two-thirds of aniridia cases are familial, inherited in an autosomal dominant manner with high penetrance, primarily due to haploinsufficiency of the *PAX6* gene on chromosome 11p13. The remaining one-third are sporadic cases caused by de novo mutations.  
  Autosomal recessive congenital aniridia is rare, accounting for 1% to 3% of cases.
* Associated Conditions:  
  Aniridia can be isolated or syndromic, occurring as part of WAGR syndrome (Wilms’ tumor, aniridia, genitourinary anomalies, intellectual disability), WAGRO syndrome (WAGR plus obesity), or Gillespie syndrome.  
  Systemic associations include diabetes, hearing difficulties, sleep disorders, and brain structural abnormalities.
* Ocular Complications:  
  Glaucoma affects approximately 46% to 70% of aniridia patients, with about 15% prevalence in children under 10 years. Cataracts and corneal opacification are also common and contribute to progressive visual impairment.
* Gender Distribution:  
  No gender predilection has been reported; males and females are equally affected

*REFERENCE:*

[Aniridia - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/sites/books/NBK538133/#article-17590.s4)

[Aniridia (Absence of Iris): What It Is, Causes & Symptoms](https://my.clevelandclinic.org/health/diseases/24116-aniridia-absence-of-iris)

**ANOPHTHALMIA AND MICROPHTHALMIA**

*ALTERNATIVE NAMES:* Microphthalmia is also known as “small eye syndrome”, and “microphthalmos”.

**DEFINITION / DESCRIPTION**

**Microphthalmia and anophthalmia**

Microphthalmia and anophthalmia are both congenital conditions that affect the eyes. A congenital condition is one that you have when you’re born. Conditions that are a result of problems with fetal development are sometimes called birth defects. You may hear some people say that anophthalmia and microphthalmia are examples of “eye birth defects.”

Microphthalmia means that one eye or both eyes don’t develop fully so they are small and disorganized. Bilateral microphthalmia is the term for when the condition affects both eyes. Unilateral microphthalmia is the term for when the condition affects only one eye.

Anophthalmia means that one or both eyes don’t develop at all so they are missing. In bilateral anophthalmia, both eyes are missing. In unilateral anophthalmia, one eye is missing.

According to some estimates, these conditions (anophthalmia and microphthalmia) affect about 1 in 5,200 to 1 in 10,000 infants born each year in the U.S.

**CAUSES**

**What causes microphthalmia and anophthalmia?**

Researchers don’t know for sure what causes anophthalmia or what causes microphthalmia. Some babies are born with these conditions due to genetic changes. Researchers think that the changes in genes and chromosomes may combine with environmental factors to result in conditions present at birth.

There are other things that may be factors in these eye conditions, including:

* Taking medications that include isotretinoin (Accutane®) or thalidomide during a pregnancy. Isotretinoin treats acne. Thalidomide treats cancer and some skin conditions.
* Being exposed to X-rays or other forms of radiation during pregnancy.
* Being exposed to chemicals, like drugs or pesticides, during pregnancy.
* Being exposed to infections like rubella and toxoplasmosis during pregnancy.
* Maternal vitamin A deficiency.

**SIGNS / SYMPTOMS**

**What are the symptoms of microphthalmia and anophthalmia?**

Your provider will be able to tell if your baby has microphthalmia or anophthalmia by looking carefully during a physical examination and doing an eye exam.

Symptoms include poor vision or even complete vision loss. These eye conditions can happen along with other eye conditions and medical issues.

Some of the other eye issues include:

* Cataracts: A cataract forms on the lens of the eye and makes it cloudy, causing poor vision and subdued colors.
* Coloboma: A coloboma means that tissue is missing in the eye. It mostly happens in the iris, or the colored part of the eye. If you have a coloboma, your pupil (the black part of the eye) may have an irregular shape because the iris has a notch or groove in it.
* Microcornea: A microcornea is a cornea that’s very small. If you have it, your cornea doesn’t reach 10 mm in diameter even when you’re an adult.
* Detached retina: This condition is serious and can cause blindness. The retina, a layer of tissue at the back of your eye, gets separated from its supporting tissue. The retina is responsible for sending signals to the brain so we can see.
* Ptosis or pseudoptosis: Ptosis refers to a drooping eyelid and involves muscles and nerves. Pseudoptosis may look like ptosis because the eyelid droops. However, the cause isn’t nerves and muscles, but an undeveloped ocular globe (eyeball).

**DIAGNOSIS METHODS**

In a newborn child, your provider can diagnose anophthalmia and microphthalmia through an examination. However, it’s also possible to diagnose these conditions during pregnancy.

Tests that can diagnose microphthalmia and anophthalmia before birth include:

Ultrasound. Ultrasonography uses high-intensity sound waves to generate images. Fetal ultrasound can’t always detect microphthalmia.

Fetal MRI. MRI stands for magnetic resonance imaging. It’s a specialized imaging test that may be helpful in evaluating for fetal congenital anomalies and associated complications.

Genetic testing, like the quad marker screening test: The quad screen is a blood test done between the 15th and 20th weeks of pregnancy that can provide information on genetic disorders in the fetus.

**TREATMENT OPTIONS**

**Management and Treatment**

Healthcare providers aren’t able to provide a new eye for people born with these conditions. However, there are treatments that include:

* Conformers: These are devices that fit into the eye socket to help your eye socket and face develop more typically. You’ll need bigger devices as your face grows.
* Prosthetic eyes: Prosthetic eyes are placed in empty eye sockets. They also help with socket and face development and can help with cosmetic concerns.
* Surgery: You might need surgery to treat cataracts, coloboma or to help with the conformer fittings.
* Services to help a child and their family deal with vision loss or blindness. Some of these specialists include teachers for the visually impaired, low vision therapists and low vision specialists. These early intervention services will help babies learn to walk, talk and interact with others. Additional services can help families work together to improve life for their child.
* Glasses or contacts. Correcting refractive error is necessary to treat any sign of amblyopia, also called lazy eye. Protective eyewear is important if you only have one eye with vision. Depending on which parts of the eye are involved in microphthalmia, you may still be able to have the ability to see clearly, especially with corrective lenses.

**PREVENTION TIPS**

There’s no way to completely eliminate your risk of microphthalmia and anophthalmia, but there are ways to make pregnancy safer:

* See a healthcare provider before you get pregnant and work together so you can be as healthy as possible before and during your pregnancy. This talk should include details on what types of vaccinations you might need to be up-to-date before you get pregnant.
* Make sure you get prenatal care (care before birth) early and consistently. Always go to your appointments, even if you feel fine.
* Talk to your provider about the medications and over-the-counter products you take to make sure that they are compatible with a healthy pregnancy. This includes prescription products and supplements. You must talk to your provider if you take isotretinoin and thalidomide.
* Your provider may suggest genetic testing before you get pregnant after discussing your medical history and your family history.
* Avoid harmful chemicals.

**How can I lower my baby’s risk of having anophthalmia and microphthalmia?**

Talk to your doctor about ways to reduce your baby’s risk of having anophthalmia and microphthalmia. Some things you can do include:

* Getting a checkup before you’re pregnant
* Talking to your doctor about medicines you take
* Visiting a genetic counselor (someone who helps you understand your genes)
* Seeing your doctor regularly for prenatal care (health care you get during pregnancy)
* Staying away from harmful things in the environment

**OUTLOOK / PROGNOSIS**

**What can I expect if I have microphthalmia or anophthalmia?**

There’s no cure for microphthalmia or anophthalmia. It’s a question of managing these conditions and any other conditions that might occur with them.

**Living With**

It’s important to have a healthcare team if you or your child has microphthalmia or anophthalmia. In addition to a pediatrician or internist, someone with either of these conditions will probably need an ophthalmologist, an ocularist and an oculoplastic surgeon.

An ophthalmologist is a medical doctor who is trained in diagnosing and treating eye conditions and vision conditions.

An ocularist is a provider who can make prosthetic devices like artificial eyes and conformers. They can also do the fitting for these devices.

An oculoplastic surgeon is a surgeon who has special training with the eyes, the eye sockets and the bones that make them up.

It’s a good idea to have all these members of your healthcare team (or your child’s team), along with experts who can help with any other areas of need

**DIFFERENTIAL DIAGNOSIS**

| **Condition** | **Distinguishing Features** |
| --- | --- |
| Anophthalmia | Complete absence of the ocular globe with presence of ocular adnexa (eyelids, conjunctiva). Confirmed by imaging showing no globe. |
| Microphthalmia | Small eye(s) with axial length ≥2 standard deviations below age norm; ocular tissue present. May be simple (isolated) or complex (with other ocular anomalies like cataracts, coloboma). |
| Cryptophthalmos | Eyelids completely fused without lashes; often associated with microphthalmia or microcornea. Usually bilateral and syndromic. |
| Cyclopia / Synophthalmia | Severe neural developmental defects with partial or complete fusion of the eyes; incompatible with life. |
| Congenital Cystic Eye | Cystic structure in orbit due to failure of optic vesicle invagination; may mimic anophthalmia at birth but cyst enlarges postnatally. |
| Congenital Cataracts | Lens opacity without globe size reduction; can coexist with microphthalmia but distinct diagnosis. |
| Coloboma | Defect in ocular structures (iris, retina) due to incomplete closure of optic fissure; may accompany microphthalmia. |
| Nanophthalmos | Small eye with microcornea, thickened sclera, hypermetropia; often considered a subtype of microphthalmia. |
| Syndromic Associations | Conditions like Aicardi syndrome, CHARGE syndrome, Lenz microphthalmia syndrome, Norrie disease, Fraser syndrome, etc., where anophthalmia/microphthalmia occur with systemic anomalies. |

*REFERENCE:* [Microphthalmia & Anophthalmia: Types, Symptoms & Treatment](https://my.clevelandclinic.org/health/diseases/24134-microphthalmia-anophthalmia)

<https://www.cdc.gov/birth-defects/about/anophthalmia-microphthalmia.html>

[Anophthalmia and Microphthalmia | National Eye Institute](https://www.nei.nih.gov/learn-about-eye-health/eye-conditions-and-diseases/anophthalmia-and-microphthalmia)

[Distinguishing risk factors between congenital anophthalmia and microphthalmia using multivariable logistic regression - PMC](https://pmc.ncbi.nlm.nih.gov/articles/PMC7327332/)

**BEHçET DISEASE**

*ALTERNATIVE NAMES:* Behcet disease is also known as “Behçet syndrome”, “Morbus Behçet (1947)”, ”Adamantiades-Behçet disease”, “Silk Road disease”, and “Gilbert-Behçet disease”.

**DEFINITION / DESCRIPTION**

Behcet (beh-CHET) disease, also called Behcet syndrome, is a rare condition. It causes blood vessel swelling, called inflammation, throughout the body.

Behcet disease symptoms can seem like they aren't related at first. They can include mouth sores, eye irritation and swelling, skin rashes and sores, and genital sores.

Treatment involves medicines to ease symptoms of Behcet disease and to prevent serious complications, such as blindness.

**CAUSES**

Behcet disease might be an autoimmune disorder, which means the body's immune system attacks some of its own healthy cells by mistake. It's likely that gene changes and factors in the environment play a role.

Experts believe that swelling of the blood vessels, called vasculitis, causes the symptoms of Behcet disease. The condition can involve arteries and veins of all sizes. Vasculitis can damage blood vessels all through the body.

Some genes might make people more likely to get Behcet disease. Some researchers think that a germ can trigger the disease in people who have these genes.

**RISK FACTORS**

Factors that might increase your risk of Behcet disease include:

* Age. Behcet disease most often affects people in their 20s and 30s. But children and older adults also can have the condition.
* Country. People from countries in the Middle East and East Asia, including Turkey, Iran, Japan and China, are more likely to get Behcet disease.
* Sex assigned at birth. Behcet disease can happen to anyone. But the disease is most often worse in people assigned male at birth.
* Genes. Certain genes may be linked with a higher risk of getting Behcet disease.

**SIGNS / SYMPTOMS**

Behcet disease symptoms vary from person to person. Symptoms can come and go or become less serious over time. Symptoms depend on which parts of the body the condition affects.

Behcet disease most often affects the following:

* Mouth. Painful mouth sores that look like canker sores are the most common sign of Behcet disease. They begin as raised, round sores in the mouth. They quickly turn into painful ulcers.  
  The sores most often heal in 1 to 3 weeks. But they often come back.
* Skin. Some people get sores that look like acne on their bodies. Others get raised and tender growths called nodules on their skin, mainly on the lower legs.
* Genitals. Open sores can happen on the scrotum or the vulva. The sores are most often painful and can leave scars.
* Eyes. Irritation and swelling, called inflammation, in the eye causes redness, pain and blurred vision. Called uveitis, this inflammation most often affects both eyes. In people with Behcet disease, these symptoms can come and go.
* Joints. Joint swelling and pain often affect the knees in people with Behcet disease. The ankles, elbows or wrists also might be involved. Symptoms can last 1 to 3 weeks and go away on their own.
* Blood vessels. Swelling, called inflammation, in veins and arteries can cause redness and pain. It can cause a blood clot, which leads to swelling in the arms or legs. Inflammation in the large arteries can lead to complications. These include bulges in the artery that can burst, called aneurysms, and narrowed or blocked blood vessels.
* Digestive system. Several symptoms can affect the network of organs that digest food, called the digestive system. Symptoms may include belly pain, diarrhea and bleeding.
* Brain. Swelling, called inflammation, in the brain and nervous system can cause headache, fever, confusion, poor balance or stroke.

**DIAGNOSIS METHODS**

No tests can show that you have Behcet disease. So your healthcare professional mainly makes the diagnosis based on your symptoms. Nearly everyone with the condition gets mouth sores. So mouth sores that come back at least three times in 12 months often are needed for a diagnosis of Behcet disease.

Also, a diagnosis of Behcet disease needs at least two other symptoms, such as:

* Genital sores that keep coming back.
* Swelling and irritation, called inflammation, of the eye.
* Skin sores.

**Tests to help diagnose Behcet disease include:**

* Lab tests. Blood tests or other lab tests might rule out other conditions.
* Pathergy test. Your healthcare professional puts a sterile needle into your skin and looks at the area 1 to 2 days later. If the test is positive, a small bump forms under your skin where the needle was put in. This shows that your immune system reacts too much to a minor injury.

**TREATMENT OPTIONS**

There's no cure for Behcet disease. If you have a mild form, your healthcare professional might suggest medicines to manage the pain and swelling of flares. You might not need medicine between flares. For some people, symptoms get better over time.

For worse symptoms, you might take medicines to manage Behcet disease throughout your body. Plus you might take medicines for flares.

**Treatments for symptoms of Behcet disease**

Medicines to manage symptoms during flares might include:

* Skin creams, gels and ointments. These products, called topical medicines, are corticosteroid medicines that you put on skin and genital sores.
* Mouth rinses. Special mouthwashes that have corticosteroids and other agents might reduce the pain of mouth sores.
* Eye drops. For mild swelling and irritation, eye drops that have corticosteroids or other medicines might help.

**Whole-body treatments for Behcet disease**

If topical medicines don't help, you might take a medicine by mouth called colchicine (Colcrys, Mitigare, others). This is for mouth and genital sores that come back. Colchicine also might ease joint swelling.

More recently, the U.S. Food and Drug Administration approved apremilast (Otezla) for the treatment of mouth sores caused by Behcet disease. Side effects may include weight loss and depression.

If you have more-serious Behcet disease, you may need treatments to lessen damage from the disease between flares. Your healthcare professional might prescribe:

* Corticosteroids to manage swelling and irritation, called inflammation. Corticosteroids, such as prednisone (Rayos), ease the inflammation caused by Behcet disease. Healthcare professionals often prescribe corticosteroids with another medicine to make the immune system less active.  
  Side effects of corticosteroids include weight gain, heartburn, high blood pressure and bone thinning, called osteoporosis.
* Medicines that make the immune system less active. Medicines that keep the immune system from attacking healthy tissues include azathioprine (Azasan, Imuran), cyclosporine (Gengraf, Neoral, others) and cyclophosphamide (Cytoxan).  
  These medicines can increase the risk of infection. They also can affect the liver and kidneys and cause low blood counts and high blood pressure.
* Medicines that change how the immune system responds. Interferon alfa-2b (Intron A) may be used alone or with other medicines to help manage skin sores, joint pain and eye irritation in people with Behcet disease. Side effects include flu-like symptoms, such as muscle pain and fatigue.  
  Medicines that block a substance called tumor necrosis factor can treat some of the symptoms of Behcet disease. People who have worse symptoms or symptoms that resist treatment might take one of these medicines.  
  Medicines include infliximab (Remicade) and adalimumab (Humira). Side effects might include headache, skin rash and a higher risk of infections.

**POSSIBLE COMPLICATIONS**

Complications of Behcet disease depend on symptoms. For instance, untreated uveitis can lead to loss of vision or blindness. If you have eye symptoms of Behcet disease, see an eye specialist, called an ophthalmologist, regularly. Treatment can help prevent this complication.

**WHEN TO SEE A DOCTOR**

Make an appointment with your healthcare professional if you have symptoms that might be Behcet disease. If you've been diagnosed with the condition, see your health professional if you get new symptoms.

**DIFFERENTIAL DIAGNOSIS**

Neurologic Behçet disease must be considered in the differential diagnosis of a variety of neurologic syndromes, including the following:

* Stroke in young adults
* Multiple sclerosis
* Movement disorders
* Intracranial hypertension
* Intracranial sinovenous occlusive diseases

Ophthalmologic considerations

It is important to consider other forms of uveitis in the differential diagnosis, especially in those patients who have a mild or atypical presentation of Behçet disease.

Human leukocyte antigen B27 (HLA-B27)–related anterior uveitis also may cause recurrent iridocyclitis with hypopyon, but it is typically unilateral.

Since Behçet disease is a bilateral panuveitis, other inflammatory processes that affect both eyes must be considered. Syphilis causes retinitis with vitritis rather than strict vasculitis. The diagnosis for syphilis is confirmed by serology.

Sarcoidosis, another bilateral inflammatory process, may produce posterior pole findings similar to

**EPIDEMIOLOGY**

**United States statistics**

Behçet disease is not common in the United States. The US prevalence is reported as 0.12-0.33 cases per 100,000 population.However, data from a study of residents of Olmsted County, Minnesota, over a 45-year period identified a prevalence of 5.2 cases per 100,000 population.

The American Southwest has a prevalence of 8.9-10.6 cases per 100,000 population, with ethnic proportions of 49.2% Hispanic American, 31.7% European-American, 14.3% Native American, and 1.7% derived from Silk Road populations.

**International statistics**

Worldwide, the incidence and prevalence of Behçet disease are highest along the Silk Road, the ancient trade passage that extended from China to the Middle East and southern Europe. This geographic pattern correlates with the prevalence of carriage of the HLA-B\*51 allele of the major histocompatibility complex, which is associated with susceptibility to Behçet disease.A meta-analysis of 18 HLA-B\*51 studies concluded that this association with Behçet disease appeared to strengthen geographically eastward along the Silk Road.

Turkey has the highest prevalence of Behçet disease, with 420 cases per 100,000 population. The prevalence in Japan, Korea, China, Iran, and Saudi Arabia ranges from 13.5 to 22 cases per 100,000 population. The prevalence in North America and Europe is much lower, with 1 case per 15,000-500,000 population.

*REFERENCES:*

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[**Behcet disease - Diagnosis and treatment - Mayo Clinic**](https://www.mayoclinic.org/diseases-conditions/behcets-disease/diagnosis-treatment/drc-20351331)

**BELL’S PALSY**

ALTERNATIVE NAMES: Bell’s palsy is also known as “acute facial palsy of unknown cause”, or simply “facial palsy”.

**DEFINITION / DESCRIPTION**

Bell’s palsy is a nerve problem that affects the muscles of your face. It causes weakness or partial paralysis of the muscles on one side of your face. With Bell’s palsy, your eyelid may not close properly and your smile may seem uneven.

Bell’s palsy usually affects adults only. It is more likely to happen to people who have diabetes, are pregnant, or have a family history of Bell’s palsy.

Bell’s palsy happens when there’s inflammation and swelling of your seventh cranial nerve — the nerve that controls facial muscles. Certain conditions (like viral infections) can cause inflammation, but many cases of Bell’s palsy have no clear cause (idiopathic).

Bell’s palsy can affect anyone at any age. But it’s most likely to affect people between the ages of 15 and 60. The average age of onset is 40 years.

The condition gets its name from Sir Charles Bell, a Scottish surgeon who first described it during the 19th century.

Is Bell’s palsy a serious condition?

Bell’s palsy isn’t a serious condition. Most cases go away on their own with time. However, the symptoms of Bell’s palsy are similar to those of serious medical conditions, like a stroke. This is why it’s important to see a healthcare provider as soon as you notice muscle weakness in your face.

Signs of a stroke include:

One-sided weakness or paralysis.

Aphasia (difficulty with or loss of speaking ability).

Loss of muscle control on one side of your face.

Sudden loss — either partial or total — of one or more senses (vision, hearing, smell, taste and touch).

Blurred or double vision (diplopia).

Loss of coordination or clumsiness (ataxia).

Dizziness.

Nausea and vomiting.

Neck stiffness.

Emotional instability and personality changes.

Seizures.

Memory loss (amnesia).

Headaches (usually sudden and severe).

A stroke is a life-threatening emergency condition where every second counts. If you or someone with you has symptoms of a stroke, Bell’s palsy is relatively common. About 15 to 30 people out of 100,000 develop it every year. About 1 in 60 people will get it at some point in their life. It’s the most common cause of one-sided facial paralysis.

**CAUSES**

Generally, it is not known what causes Bell’s palsy. However, doctors believe it may be due to one or more of these problems:

* problems in your body’s immune system (how it fights disease)
* reduced blood flow to a nerve that goes to your face (the 7th cranial nerve)
* infection from a virus, causing swelling of the facial nerves

Although the exact reason Bell's palsy occurs isn't clear, it's often related to having a viral infection. Viruses that have been linked to Bell's palsy include viruses that cause:

* Cold sores and genital herpes, also known as herpes simplex.
* Chickenpox and shingles, also known as herpes zoster.
* Infectious mononucleosis, caused by the Epstein-Barr virus.
* Cytomegalovirus infections.
* Respiratory illnesses, caused by adenoviruses.
* German measles, also known as rubella.
* Mumps, caused by the mumps virus.
* Flu, also known as influenza B.
* Hand-foot-and-mouth disease, caused by a coxsackievirus.

The nerve that controls facial muscles passes through a narrow corridor of bone on its way to the face. In Bell's palsy, that nerve becomes inflamed and swollen — usually related to a viral infection. Besides affecting facial muscles, the nerve affects tears, saliva, taste and a small bone in the middle of the ear.

**RISK FACTORS**

Bell's palsy occurs more often in people who:

* Are pregnant, especially during the third trimester, or who are in the first week after giving birth.
* Have an upper respiratory infection, such as the flu or a cold.
* Have diabetes.
* Have high blood pressure.
* Have obesity.

It's rare for Bell's palsy to come back. But when it does, there's often a family history of repeated attacks. This suggests that Bell's palsy might have something to do with genes.

**SIGNS / SYMPTOMS**

Symptoms of Bell's palsy come on suddenly and may include:

* Mild weakness to total paralysis on one side of the face — occurring within hours to days.
* Facial droop and trouble making facial expressions, such as closing an eye or smiling.
* Drooling.
* Pain around the jaw or pain in or behind the ear on the affected side.
* Increased sensitivity to sound on the affected side.
* Headache.
* Loss of taste.
* Changes in the amount of tears and saliva produced.

Rarely, Bell's palsy can affect the nerves on both sides of the face.

Your eyes may become dry and you may have blurry vision. One eye may not close completely, and it may feel irritated.

You might not be able to taste food as well as you could before. Also, you could have hearing problems, such as having things sound distorted or unusual.

Tell your primary care doctor or ophthalmologist if any of your symptoms get worse.

If you have symptoms on **both** sides of your face, you may have something other than Bell’s palsy. If your symptoms do not improve in a few weeks, your ophthalmologist may recommend an MRI. An MRI is a scan that provides images of tissue inside the body.

**How long do Bell’s palsy symptoms last?**

For about 8 out of 10 people, symptoms of Bell’s palsy start to improve in about 3 weeks. Symptoms should be nearly gone in about 2 to 3 months.

Some symptoms may remain, such as a small amount of facial paralysis or reduced movement on one side of your face. For about 2 out of 10 people, Bell’s palsy symptoms never go away.

Your ophthalmologist can tell you how to relieve uncomfortable eye-related symptoms from Bell’s palsy.

**DIAGNOSIS METHOD**

There's no specific test for Bell's palsy. Your healthcare professional looks at your face and asks you to move your facial muscles. You're asked to close your eyes, lift your brow, show your teeth and frown, among making other movements.

Other conditions — such as a stroke, infections, Lyme disease, inflammatory conditions and tumors — can cause facial muscle weakness that mimics Bell's palsy. If the cause of your symptoms isn't clear, your healthcare professional may recommend other tests, including:

* **Electromyography (EMG).** This test can confirm the presence of nerve damage and determine how serious it is. An EMG measures the electrical activity of a muscle in response to stimulation. It also measures the nature and speed of the conduction of electrical impulses along a nerve.
* **Imaging scans.** Magnetic resonance imaging (MRI) or computerized tomography (CT) may be needed on occasion to rule out other possible sources of pressure on the facial nerve, such as a tumor or skull fracture.
* **Blood tests.** There is no blood test for Bell's palsy. But blood tests can be used to rule out Lyme disease and other infections.

**TREATMENT OPTIONS**

There is no main treatment for Bell’s palsy. In most cases, it goes away on its own in a few weeks. However, your ophthalmologist can help manage the symptoms affecting your eye. Eye drops or other lubricants provide relief if you cannot fully shut your eye.

In some cases, corticosteroids, antiviral drugs or other medicine may be prescribed to help you heal from Bell’s palsy

Most people with Bell's palsy recover fully — with or without treatment. There's no one-size-fits-all treatment for Bell's palsy. But your healthcare professional may suggest medicines or physical therapy to help speed your recovery. Surgery is rarely an option for Bell's palsy.

Because the eye on the affected side doesn't close, it's important to take steps to protect and care for that eye. Use lubricating eye drops during the day and an eye ointment at night to help keep your eye moist. Wear glasses or goggles during the day and an eye patch at night to protect your eye from getting poked or scratched. You may need to see a healthcare professional to monitor your eye.

**MEDICINES**

Commonly used medicines to treat Bell's palsy include:

* **Corticosteroids,** such as prednisone (Rayos, Prednisone Intensol). These are powerful anti-inflammatory agents. If they can reduce the swelling of the facial nerve, the nerve can fit more comfortably within the bony corridor that surrounds it. Corticosteroids may work best if they're started within several days of when symptoms start. Steroids started early improve the likelihood of complete recovery.
* **Antiviral drugs.** The role of antivirals is not certain. Antivirals alone have shown no benefit compared with placebo. Antivirals added to steroids may benefit some people with Bell's palsy, but this is still not proved.

Despite this, an antiviral medicine, such as valacyclovir (Valtrex) or acyclovir, is sometimes given in combination with prednisone in people with severe facial palsy.

**Physical therapy**

Paralyzed muscles can shrink and shorten, which may be permanent. A physical therapist can teach you how to massage and exercise your facial muscles to help prevent this from occurring.

**Surgery**

In the past, decompression surgery was used to relieve the pressure on the facial nerve by opening the bony passage that the nerve passes through. Today, decompression surgery isn't recommended. Facial nerve injury and permanent hearing loss are possible risks associated with this surgery.

Rarely, plastic surgery may be needed to correct lasting facial nerve problems. Facial reanimation surgery helps make the face look more even and may restore facial movement. Examples of this type of surgery include an eyebrow lift, an eyelid lift, facial implants and nerve grafts. Some procedures, such as an eyebrow lift, may need to be repeated after several years.

**POSSIBLE COMPLICATIONS**

Mild symptoms of Bell's palsy typically disappear within a month. Recovery from more-complete facial paralysis can vary. Complications may include:

* Irreversible damage to your facial nerve.
* Irregular regrowth of nerve fibers. This may result in involuntary contraction of certain muscles when you're trying to move other muscles, known as synkinesis. For example, when you smile, the eye on the affected side may close.
* Partial or complete blindness of the eye that won't close. This is caused by excessive dryness and scratching of the clear protective covering of the eye, known as the cornea.

**WHEN TO SEE A DOCTOR**

Seek medical help right away if you experience any type of paralysis because you may be having a stroke. Bell's palsy is not caused by a stroke, but the symptoms of both conditions are similar.

If you have facial weakness or drooping, see your healthcare professional to find out the cause and the severity of the illness.

**Lifestyle and home remedies**

Home treatment may include:

* **Taking pain relievers.** Aspirin, ibuprofen (Advil, Motrin IB, others) or acetaminophen (Tylenol, others) are available without a prescription and may help ease your pain.
* **Doing physical therapy exercises.** Massaging and exercising your face according to your physical therapist's advice may help relax your facial muscles.

**Alternative medicine**

Although there's little scientific evidence to support the use of alternative medicine to treat Bell's palsy, some people with the condition may benefit from the following:

* **Acupuncture.** Placing thin needles into a specific point in the skin helps stimulate nerves and muscles, which may offer some relief.
* **Biofeedback training.** By teaching you to use your thoughts to control your body, biofeedback training may help you gain better control over your facial muscles.
* **OnabotulinumtoxinA (Botox).** This medicine may help manage symptoms such as facial spasms and tearing. Injections of onabotulinumtoxinA also may help improve the balance of the face

**DIFFERENTIAL DIAGNOSIS**

## Central (Upper Motor Neuron) Causes

* Stroke (Cerebrovascular Accident)
  + Typically spares the forehead due to bilateral cortical innervation.
  + Often accompanied by other neurological deficits (weakness, numbness).
* Multiple Sclerosis
* Brain Tumors or Mass Lesions
* Subdural Hematoma

*Key clinical clue*: Forehead sparing and additional neurological signs suggest central cause.

2. Infectious Causes

* Herpes Simplex Virus-1 (HSV-1) (most common presumed cause of Bell palsy)
* Herpes Zoster Virus (Ramsay Hunt Syndrome)
  + Facial palsy with vesicular rash in ear canal, external ear, or oropharynx.
  + Severe pain often precedes palsy.
* Lyme Disease
  + History of tick exposure, erythema migrans rash, arthralgia.
  + Often bilateral facial palsy.
* Acute Otitis Media or Cholesteatoma
  + Gradual onset, ear pain, fever, conductive hearing loss.
* Other infections: HIV, syphilis, tuberculosis, sarcoidosis (neurosarcoidosis).

3. Neoplastic Causes

* Parotid Gland Tumors
* Cerebellopontine Angle Tumors (e.g., acoustic neuroma, meningioma)
* Metastatic Lesions
* Leukemic or Lymphomatous Infiltration

These usually cause gradual onset and progressive symptoms.

4. Traumatic Causes

* Temporal bone fractures
* Iatrogenic injury (e.g., surgery involving parotid gland or mastoid)

5. Other Causes

* Sarcoidosis (Heerfordt syndrome with facial palsy)
* Guillain-Barré Syndrome (Miller Fisher variant)
* Diabetes Mellitus (may predispose to facial nerve palsy)
* Idiopathic (Bell Palsy)

Clinical Features to Differentiate Bell Palsy

* Lower Motor Neuron Pattern: Involves entire half of face including forehead.
* Sudden onset: Usually over hours to days.
* Absence of other neurological signs: No limb weakness or sensory changes.
* Associated symptoms: Hyperacusis, altered taste, reduced lacrimation may be present.

**EPIDEMIOLOGY**

The annual incidence of Bell palsy is 15 to 40 per 100,000 individuals, and the lifetime risk is 1 in 60, with a recurrence rate of 8% to 12%. There is no sex, ethnic, or laterality predilection, and Bell palsy can occur at any age; there is a bimodal distribution with incidence peaks between 20 and 30 years and between 60 and 70. There are multiple known risk factors for developing Bell palsy, including diabetes, pregnancy, preeclampsia, obesity, dental procedures, and, debatably, hypertension. Pregnant patients and those with diabetes are specifically at higher risk for worse outcomes and are potentially more likely to present with worse paralysis than patients without diabetes and who aren't pregnant with Bell palsy.

According to a 2022 paper published by Escalante et al, the most common severity of Bell palsy is House-Brackmann grade III (mild-moderate), accounting for 41.9% of patients. Grade VI palsy (total hemifacial paralysis) occurs in 20.1% of patients. House-Brackmann grades II (mild) and V (severe) each comprise 16.3% of patients, and grade IV accounts for only 5.4%. The same study also assessed the chance of recovery as a function of palsy severity and found that patients with House-Brackmann grade VI palsy had a 60% chance of recovery to grade I or II, and grade V patients had an 83% chance if provided steroids and antivirals. Patients with grade II to IV paralysis all recovered to grade I or II in this series.

*REFERENCES:*

[Bell Palsy - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK482290/)

[Bell's palsy - Symptoms and causes - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/bells-palsy/symptoms-causes/syc-20370028)

[Bell's palsy - Diagnosis and treatment - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/bells-palsy/diagnosis-treatment/drc-20370034)

**BEST DISEASE (BEST VITELLIFORM MACULAR DYSTROPHY)**

ALTERNATIVE NAMES: Best vitelliform macular dystrophy is also known as “vitelliform macular dystrophy 2 (Best disease, bestrophin)”, “Best Disease”, “BEST”, “BEST1\_HUMAN”, “BMD”, and “VMD”.

**DEFINITION / DESCRIPTION**

Best disease is an inherited disease that affects the retinas of your eyes. It causes the macula, which is the central part of the retina, to degrade. This means that you can have problems with your central vision, or seeing things that are right in front of you. Best disease may not affect peripheral (side) vision.

Other names for Best disease include vitelliform macular dystrophy or vitelliform dystrophy. Dystrophy is the medical name for the degeneration of an organ.

There’s a form of vitelliform macular dystrophy that doesn’t start when you’re young but instead happens when you get older, usually from 40 to 60 years of age. This is the adult-onset type of the disease.

It’s not clear how many people actually inherit Best disease. One study indicates that Best disease happens in an estimated 1 in 16,500 to 1 in 21,000 people. Another estimate puts the figure at about 1 in 15,000 people and yet another puts the figure at 1 in 10,000 people.

**Stages of Best disease**

Best disease can go through six stages. They are:

* Stage I - Pre Vitelliform: You probably don’t have symptoms. You haven’t yet developed any of the yellow material underneath your retina.
* Stage II - Vitelliform: This word (vitelliform) means “shaped like an egg.” At this stage, the yellow material is collected in an egg-like shape. Your vision may still be fine.
* Stage III - Pseudohypopyon: The yellow material may develop into a cyst under the retina.
* Stage IV - Vitelliruptive: The material may begin to damage the cells of the retina and you may notice changes in vision.
* Stage V- Atrophic: The yellow material goes away, but it leaves scars and damaged cells. Some researchers consider this to be the final stage of Best disease.
* Stage VI - Choroidal neovascularization or CNV: Some researchers consider CNV to be the final stage of Best disease, while others think that CNV is a complication of the disease. CNV happens in about 20% of people who have Best disease. Neovascularization refers to new blood vessels that grow in the choroid. The choroid is the layer of tissue between the retina and the sclera (the white part of your eye). Those blood vessels may leak and cause your vision to get worse.

**CAUSES**

A genetic condition causes Best disease.

Best disease is an autosomal dominant condition. This means that only one parent needs to have an altered gene to pass it on. It also means that half of the children of a parent with an autosomal trait will get that particular trait. In this case, the trait is Best disease.

The gene that’s affected in Best disease is the gene that can be affected in some types of retinitis pigmentosa, a group of inherited eye diseases.

**RISK FACTORS**

Since vitelliform macular dystrophy is hereditary, there are **no risk factors for developing the disorder other than genetics**. The only people at risk have a parent who has the mutations in the BEST1 gene.

**SIGNS / SYMPTOMS**

What are the symptoms of Best disease?

You’ll probably find out about Best disease after an eye exam. Sometimes there are no signs and symptoms. When there are, signs and symptoms may include:

* Blurred vision.
* Problems with central vision but not with side vision.
* Seeing things that appear to be shaped oddly, like wavy lines instead of straight lines. This is called metamorphopsia.
* Severe vision loss.
* Different levels of vision or vision loss in each eye.

**DIAGNOSIS METHODS**

In many cases, a healthcare provider will find Best disease when you’re between five and 10 years old or by the time you’re 20 years old.

An eye care specialist will first take a medical history and do an eye exam. They may also ask for imaging tests and measure how thick your choroid is. Tests may include:

* Fluorescein angiography: This is an imaging test that uses dye to show blood vessels.
* Optical coherence tomography: This type of noninvasive imaging test uses reflected light to create pictures of the back of your eye.
* Color fundus photography: This type of imaging shows your retina, the related blood vessels and the optic nerve head.
* Ophthalmic electrophysiology: This term describes a series of eye tests that measure electrical activity.
* Genetic tests: These tests will give the provider information about your genes.

The eye care provider may also want to speak to family members or examine their eyes.

**TREATMENT OPTIONS**

There’s no cure for Best disease at this time.

Managing Best disease depends on the symptoms and stage of the condition. Once it’s diagnosed, you’ll need to keep a regular schedule of eye appointments so any changes can be found early.

In early stages, you might not need any treatment. You may need treatment for other conditions, such as corrective lenses for refractive errors. People with Best disease often have farsightedness. If you have cataracts, you may need cataract surgery.

If you have choroid neovascularization and you’ve developed a choroid neovascularization membrane, the provider may suggest the following to stop the growth of new blood vessels:

* Anti-vascular endothelial growth factor agents: These are medications injected into the vitreous cavity of your eye. They’re intended to stop new blood vessels from growing.
* Laser therapy: This treatment uses lasers to close off the blood vessels.
* Photodynamic therapy: This treatment combines a laser and a drug that’s activated by light to damage the blood vessels and stop leaking.

Your provider may also suggest you use low vision aids like magnifying devices and other helpful tools.

Researchers are working on treatments or preventatives that use genetic material. Gene therapy is experimental.

**PREVENTION TIPS**

How can Best Vitelliform Macular Dystrophy be Prevented? Currently, Best Vitelliform Macular Dystrophy may not be preventable, since it is a genetic disorder.

* Genetic testing of the expecting parents (and related family members) and prenatal diagnosis (molecular testing of the fetus during pregnancy) may help in understanding the risks better during pregnancy
* If there is a family history of the condition, then genetic counseling will help assess risks, before planning for a child
* Active research is currently being performed to explore the possibilities for treatment and prevention of inherited and acquired genetic disorders

**Regular medical screening at periodic intervals with tests and physical examinations** are recommended

**How can I reduce my risk of developing Best disease?**

You can’t stop yourself from developing Best disease. If you have the genetic background to pass on Best disease, you may be interested in genetic counseling.

**OUTLOOK / PROGNOSIS**

Best disease isn’t fatal, but it’s not curable, either. You won’t go blind from Best disease, but you may have low vision.

**Living With**

**How do I take care of myself?**

Certain types of foods may help with eye health. Consider eating fish a few times per week and consuming leafy green vegetables along with other types of fruits and vegetables and nuts.

If you have Best disease, you should have regular healthcare appointments, including regular eye care appointments. Let the care provider know about any changes in your vision or your health. You may want to ask questions like:

* Can you recommend a genetic counselor to help me decide about having children?
* Am I eligible to participate in a clinical trial?
* Can you recommend an optometrist that specializes in low vision?
* Is gene therapy an option for treatment?
* The prognosis of Best Vitelliform Macular Dystrophy is dependent upon the severity of the signs and symptoms and associated complications, if any
* Individuals with mild conditions have better prognosis than those with severe symptoms and complications
* Typically, the prognosis may be assessed on a case-by-case basis

**DIFFERENTIAL DIAGNOSIS**

* Adult-Onset Vitelliform Macular Dystrophy (Pattern Dystrophy)
* Autosomal-Recessive Bestrophinopathy
* Basal Laminar Drusen
* Central Serous Chorioretinopathy With Subretinal Fibrin
* Exudative (Wet) Age-Related Macular Degeneration (AMD)
* Fundus Flavimaculatus (Stargardt Disease)
* Resolving Subretinal Hematoma

**EPIDEMIOLOGY**

*FREQUENCY*

**United States**

Best disease is rare.

**International**

Best disease is rare.

**Mortality/Morbidity**

Visual acuity is good in the pre vitelliform stage. Even with the egg-yolk appearance, visual acuity is maintained in the range of 20/20 to 20/50 (6/6 to 6/15) for many years. The breakup of the vitelliform stage, leading to the scrambled egg stage, may be accompanied by visual acuity deterioration. It is the final stage of geographic RPE atrophy with possible development of a choroidal neovascular membrane that is associated with further deterioration in visual acuity.These changes usually occur in individuals older than 40 years. Various studies have shown that most individuals retain reading and driving vision in at least 1 eye into adulthood (88% have 20/40 or better vision). Only 4% of these individuals develop vision less than 20/200 in the better eye.

**Race**

Best disease is most common in individuals of European ancestry but can also be found in individuals of African and Hispanic ancestry.

**Sex**

No known gender predilection exists.

**Age**

Usual onset of Best disease is from 3-15 years, with an average age of 6 years. The condition often is not detected until much later in the disease because visual acuity may remain good for many years. The atrophic stage usually occurs after age 40 years.

REFERENCE : [**Best Disease Differential Diagnoses**](https://emedicine.medscape.com/article/1227128-differential?form=fpf)

[**Best Disease: Background, Pathophysiology, Epidemiology**](https://emedicine.medscape.com/article/1227128-overview#a6)

[**Best Disease (Vitelliform Macular Dystrophy): Stages & Symptoms**](https://my.clevelandclinic.org/health/diseases/24132-best-disease)

[**Best Vitelliform Macular Dystrophy - DoveMed**](https://www.dovemed.com/diseases-conditions/best-vitelliform-macular-dystrophy#:~:text=How%20can%20Best%20Vitelliform%20Macular%20Dystrophy%20be%20Prevented?,intervals%20with%20tests%20and%20physical%20examinations%20are%20recommended.)

<http://www.dovemed.com/diseases-conditions/rare-disorders/>

**BIRDSHOT CHORIORETINOPATHY**

*ALTERNATIVE NAMES:* Birdshot chorioretinopathy is also known as “Birdshot chorioretinitis”, “Birdshot retinochoroiditis”, “Birdshot retinochoroidopathy”, “Birdshot uveitis”, “BSCR”, “Posterior uveitis”, and “Vitiliginous choroiditis”.

**DEFINITION / DESCRIPTION**

Birdshot chorioretinopathy (koor-ee-oh-reht-in-OHP-ah-thee), or BSCR, is a rare inflammatory eye disease. It’s a long-term (chronic) condition that can lead to permanent vision changes or complete loss of sight.

BSCR is a severe form of uveitis, a group of diseases that damage eye tissue. It causes swelling of the:

* Retina, the light-sensitive tissue at the back of your eye.
* Choroid, which connects your retina to the white part of your eye.

Hallmark signs of birdshot chorioretinopathy are cream-colored oval spots that are present in the deep retina and superficial (top layer) choroid. Early symptoms often involve floaters, blurred vision or both. The condition usually affects both eyes.

Over time, damage can cause cystoid macular edema or glaucoma. While there's no cure, birdshot chorioretinopathy is a treatable disease. Early detection is essential to the success of the treatment.

Anyone can develop birdshot chorioretinopathy. It most often affects middle-aged, white adults between the ages 40 and 60. It rarely affects children.

**Are there other conditions like birdshot chorioretinopathy?**

Several conditions, including other forms of uveitis, have symptoms that resemble birdshot chorioretinopathy. Getting an accurate diagnosis is crucial so you can get appropriate treatment. Conditions similar to BSCR include:

* Lymphoma.
* Sarcoidosis.
* Sympathetic ophthalmia, a rare type of uveitis caused by trauma to your eye.
* Syphilis.
* Presumed ocular histoplasmosis syndrome (POHS).
* Tuberculosis.
* Vogt-Koyanagi-Harada syndrome, a nervous system disease that affects vision and hearing.

A group of inflammatory conditions known as white dot syndromes also share symptoms of BSCR. They cause damage (lesions) to the retina and choroid and include:

* Acute posterior multifocal placoid pigment epitheliopathy (APMPPE).
* Multifocal choroiditis and panuveitis (MCP).
* Multiple evanescent white dot syndrome (MEWDS).
* Punctate inner choroidopathy (PIC).

**Why is it called birdshot chorioretinopathy?**

Birdshot chorioretinopathy is a relatively new disease, first described in 1949. In 1980, two eye specialists named it “birdshot chorioretinopathy” for its unique cream and orange oval-shaped spots scattered throughout the retina. The spots resemble the pattern from birdshot, small shotgun pellets (bullets).

Other names for birdshot chorioretinopathy are:

* Birdshot chorioretinitis.
* Birdshot retinochoroiditis.
* Birdshot retinochoroidopathy.
* Birdshot uveitis.
* BSCR.
* Posterior uveitis.
* Vitiliginous choroiditis.

**CAUSES**

Researchers don’t know the exact cause of birdshot chorioretinopathy. They suspect it may be an autoimmune disease, when your body's immune system mistakenly attacks healthy tissues.

More than 90% of people diagnosed with BSCR inherit (receive from their parents) a substance (antigen) called HLA-A29 that triggers an immune response. However, you don't need to have HLA-A29 to develop birdshot chorioretinopathy. You can also have the antigen and not get the disease.

**SIGNS / SYMPTOMS**

Symptoms of birdshot chorioretinopathy can vary. It's a progressive disease with symptoms that typically worsen over several months to years.

The first signs of birdshot chorioretinopathy are often floaters or blurred vision. BSCR does n't usually involve eye pain or redness. You may also experience:

* Changing vision or vision loss.
* Decreased ability to see to the side (peripheral vision) or judge depth.
* High eye pressure (ocular hypertension).
* Night blindness (nyctalopia).
* Problems with color vision (color blindness) and telling certain colors apart (dyschromatopsia).
* Sensitivity to bright lights or glare (photophobia).

Some people experience unusual changes in vision or perception. Symptoms may include:

* Ceiling fan effect (when you close your eyes, you see something like a ceiling fan or pinwheel whirring around).
* Seeing flashing or flickering lights (photopsia) or distorted shapes (metamorphopsia).
* Shimmering blurred or hazy vision (like looking through water or dirty glass).

In severe cases, symptoms may include cataracts or glaucoma, which can cause severe vision loss even after inflammation is treated. Birdshot chorioretinopathy may also lead to issues with the central retina, including cystoid macular edema and epiretinal membranes.

**DIAGNOSIS METHODS**

Diagnosis of birdshot chorioretinopathy can be difficult. It's a rare disease, and its unique spots may not be visible in early stages of the disease.

Eye care specialists (ophthalmologists) most often diagnose BSCR with an eye exam. They also use a blood test to confirm that you have the HLA-A29 antigen.

Healthcare providers may use a combination of tests to monitor you over time. Stay in touch with your provider to receive the care you need.

**TREATMENT OPTIONS**

*Management and Treatment*

There's no cure for birdshot chorioretinopathy. But with timely diagnosis, treatment can reduce symptoms and prevent worsening of the disease.

Who might be on my treatment team for birdshot chorioretinopathy?

Your care team recommends treatment, monitors you and adjusts medications based on your condition and any side effects. Your team may include:

* Eye specialist (ophthalmologist).
* Kidney (renal) specialist (nephrologist).
* Eye specialist with special training in immune disorders (ocular immunologist).
* Inflammatory disease specialist (rheumatologist).
* Musculoskeletal specialist (orthopaedist).

**How is birdshot chorioretinopathy treated?**

Treatment for birdshot chorioretinopathy depends on the severity of your condition and how it changes over time. The goal of treatment is to reduce swelling in your eyes and prevent or reverse vision loss.

Healthcare providers typically first recommend high doses of corticosteroids to control inflammation in your eye. They then lower the dose as much as possible due to the risk of side effects. You may receive steroids through:

* Eye drops.
* Oral medicines.
* Implant placed surgically in your eye (intravitreal implant).
* Injection.
* IV (intravenously, through a vein).

Many people also need long-term treatment with immunosuppressants. These drugs stop your immune system from attacking your eyes and may include one or more:

* Alkylating agents.
* Antimetabolite drugs.
* Biologic agents.
* T-cell transduction/calcineurin inhibitors.

**Are there any side effects from birdshot chorioretinopathy treatment?**

Long-term steroid and immunosuppressant use can cause serious side effects, including gastrointestinal or bone issues. They may also increase your risk of developing cataracts or glaucoma.

Your healthcare provider will monitor you for any side effects and prescribe other medications if needed. If you're taking steroids by mouth, providers will check you for bone damage (osteoporosis) and fracture risk.

Your provider may also recommend a bone density scan. You may receive medication to prevent bone loss (bisphosphonates).

Watch your condition for any changes or complications. Your healthcare provider will check your blood and urine to make sure your kidneys, liver and bone marrow are working as they should. You are also likely to have regular eye exams, including visual field tests and optical coherence tomography (OCT) scans.

**PREVENTION TIPS**

There's no known way to prevent birdshot chorioretinopathy. In most cases, it's believed to be triggered by a specific antigen (a molecule that triggers an immune response) passed down in families.

Regular eye exams can help detect early symptoms, so you can receive timely treatment if needed. Tell your healthcare provider if any family members have eye disease.

**OUTLOOK / PROGNOSIS**

The outlook for birdshot chorioretinopathy varies depending on the severity of the condition and the effectiveness of treatment. In milder cases, vision can remain stable or improve with treatment.

**Living With**

Birdshot chorioretinopathy requires prompt diagnosis and treatment from an ophthalmologist. Even if your vision is fine, an eye exam can detect low levels of inflammation.

Alert your healthcare provider to any side effects of medication or change in symptoms or vision. You may receive different medications as needed. Your healthcare provider can discuss options that work best for you.

**DIFFERENTIAL dIAGNOSIS**

*Infectious*

* Tuberculosis
* Syphilis
* Ocular histoplasmosis syndrome

*Non-infectious*

* Sarcoidosis
* Vogt-Koyanagi-Harada syndrome
* Sympathetic ophthalmia
* Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)
* Multiple evanescent white dot syndrome (MEWDS)
* Multifocal choroiditis and panuveitis (MCP)
* Punctate inner choroidopathy (PIC)

Lymphoma can masquerade as birdshot retinopathy.

**EPIDEMIOLOGY**

Birdshot chorioretinopathy is an unusual cause of ocular inflammation, and obtaining reliable incidence and prevalence data is difficult due to the rarity of the disease. Studies from Europe and the United States report that BCR accounts for between 0.5% and 1.5% of the uveitis cases seen in specialist uveitis practices. BCR is predominantly seen in the middle-aged, and some reports appear to be more common in females. In a systematic review from 2005, Shah et al. reported a mean age of disease onset of 53.0 years (512 patients), and a 54.1 % female preponderance (522 patients). Birdshot chorioretinopathy is most prevalent in White populations, is most commonly diagnosed in people of Northern European ancestry, with only the occasional case report of BCR in Latino-Hispanic, African-American, and Japanese people. The ethnic distribution is relevant to HLA-A29. HLA-A29 subtypes can be observed in many patients, with each subtype having a different prevalence.

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**BLEPHARITIS**

*ALTERNATIVE NAMES:* Blepharitis is also known as “granulated eyelids”, or “eye dandruff”.

**DEFINITION / DESCRIPTION**

Blepharitis is usually caused by an inflammation of the eyelids margin which causes redness, itching, and crusting. They may appear red, swollen, or feel like they are burning or sore. You may have flakes or oily particles (crusts) wrapped at the base of your eyelashes too. Blepharitis is very common, especially among people who have oily skin, dandruff or rosacea.

Blepharitis (blef-uh-RYE-tis) is inflammation of the eyelids.

Blepharitis commonly occurs when tiny oil glands near the base of the eyelashes become clogged, causing irritation and redness. Several diseases and conditions can cause blepharitis.

Blepharitis is often a chronic condition that's difficult to treat. Blepharitis can be uncomfortable and unsightly. But it usually doesn't cause permanent damage to your eyesight, and it's not contagious.

**Types of blepharitis**

There are two types of blepharitis, depending on where it’s located on your eyelids. They are:

* **Anterior blepharitis:** This type occurs when your eyelid’s front exterior, where the eyelashes come out of your lids, is red or darker in color and swollen, or when you have dandruff on your lashes.
* **Posterior blepharitis:** This type happens when the oil-producing meibomian glands under your eyelid produce thickened/unhealthy oil.

**CAUSES**

The exact cause of blepharitis isn't clear. It might be associated with one or more of the following:

* Seborrheic dermatitis — dandruff of the scalp and eyebrows
* Infection
* Clogged or malfunctioning oil glands in your eyelids
* Rosacea — a skin condition characterized by facial redness
* Allergies (including allergic reactions to eye medications, contact lens solutions or eye makeup)
* Eyelash mites or lice
* Dry eyes

**RISK FACTORS**

In addition to having rosacea and dandruff, you may be more at risk of developing blepharitis if you:

* Have diabetes.
* Wear contact lenses.
* Are exposed to irritants like dust and chemicals.
* Work or live in dry environments. This includes spending a lot of time in air conditioning.
* Have a high number of microbes that normally live on your skin.
* Don’t remove makeup thoroughly.
* Have oily skin.
* Are on certain drugs such as those for cancer treatment.
* Are going through menopause or hormonal changes.

Poor hygiene can be a factor in blepharitis, but it’s not as simple as saying that only people with poor hygiene can get blepharitis. Hygiene is just a part of the reason that some people get blepharitis.

If you think about it, most people don’t clean their eyelids and lashes every day or night. However, people with risk factors may have to make eyelid and eyelash hygiene a priority.

**SIGNS / SYMPTOMS**

Blepharitis signs and symptoms are typically worse in the morning. They include:

* Watery eyes
* Red eyes
* A gritty, burning or stinging sensation in the eyes
* Eyelids that appear greasy
* Itchy eyelids
* Red, swollen eyelids
* Flaking of the skin around the eyes
* Crusted eyelashes
* Eyelid sticking
* More frequent blinking
* Sensitivity to light
* Blurred vision that usually improves with blinking

**DIAGNOSIS METHODS**

Tests and procedures used to diagnose blepharitis include:

* **Examining your eyes.** Your doctor might use a special magnifying instrument to examine your eyelids and your eyes.
* **Swabbing skin for testing.** In certain cases, your doctor might use a swab to collect a sample of the oil or crust that forms on your eyelid. This sample can be analyzed for bacteria, fungi or evidence of an allergy.

**TREATMENT OPTIONS**

Self-care measures, such as washing your eyes and using warm compresses, might be all that's needed for most cases of blepharitis. If self-care measures aren't enough, your doctor might suggest prescription treatments, including:

* **Medications that fight infection.** Antibiotics applied to the eyelid have been shown to provide relief of symptoms and resolve bacterial infection of the eyelids. These are available in several forms, including eye drops, creams and ointments.

If you don't respond to topical antibiotics, your doctor might suggest an oral antibiotic.

* **Medications to control inflammation.** Steroid eye drops or ointments are used for this, generally only for people who don't respond to other therapies. Your doctor might prescribe both antibiotic and anti-inflammatory drugs.
* **Medications that affect the immune system.** Topical cyclosporine (Restasis) has been shown to offer relief of some signs and symptoms of blepharitis.
* **Treatments for underlying conditions.** Blepharitis caused by seborrheic dermatitis, rosacea or other diseases might be controlled by treating the underlying disease.

Other treatment options, such as using intense pulsed light might unclog the glands. More study is needed.

Blepharitis rarely disappears completely. Even with successful treatment, the condition frequently is chronic and requires daily attention with eyelid scrubs. If you don't respond to treatment, or if you've also lost eyelashes or only one eye is affected, the condition could be caused by a localized eyelid cancer

**Self-care**

Self-care measures might be the only treatment needed for most cases of blepharitis.

**Clean your eyes daily**

If you have blepharitis, follow this self-care remedy two to four times a day during flare-ups and once or twice a day after the condition is under control:

* Apply a warm compress over your closed eye for a few minutes to loosen the crusty deposits on your eyelids.
* Firmly but gently massage the eyelids, using a clean washcloth or a clean finger.
* Immediately use a clean washcloth or cotton-tipped applicator moistened with warm water and a few drops of diluted baby shampoo or an over-the-counter eyelid cleanser to wash away oily debris or scales at the base of your eyelashes. Use a different clean cloth for each eye.
* In some cases, you might need to be more deliberate about cleaning the edge of your eyelids at your eyelashes. To do this, gently pull your eyelid away from your eye and use the washcloth to gently rub the base of the lashes. This helps avoid damaging your cornea with the washcloth.

Ask your doctor whether you should use a topical antibiotic ointment after cleaning your eyelids in this way.

* Rinse your eyelids with warm water and gently pat them dry with a clean, dry towel.

It might help to stop using eye makeup when your eyelids are inflamed. Makeup can make it harder to keep your eyelids clean and free of debris. Also, it's possible that makeup could reintroduce bacteria to the area or cause an allergic reaction.

**Lubricate your eyes**

Try over-the-counter artificial tears. These eye drops can help relieve dry eyes.

**Control dandruff and mites**

If you have dandruff that's contributing to your blepharitis, ask your doctor to recommend a dandruff shampoo. Using a dandruff shampoo might relieve your blepharitis signs and symptoms.

Using tea tree oil shampoo on your eyelids daily might help deal with mites. Or try gently scrubbing your lids once a week with 50% tea tree oil, which is available over-the-counter. Contact your doctor if you don't see improvement in six weeks. And stop using tea tree oil if it irritates your skin or eyes.

**Alternative medicine**

No alternative medicine treatments have been proved to ease the symptoms of blepharitis. However, a diet rich in omega-3 fatty acids or supplements containing omega-3 fatty acids might help blepharitis associated with rosacea. Omega-3 fatty acids are found in foods such as salmon, tuna, trout, flaxseed and walnuts. More study is needed.

**Keeping Blepharitis Under Control**

It is very important to keep your eyelids, skin and hair clean. This will help keep your blepharitis symptoms under control. Use baby shampoo diluted in warm water to gently scrub the eyelids/eyelashes daily when you have a crusting present.

There are also wipes and some antiseptic sprays you can use to scrub to keep bacteria from growing too much. Scrub for about 15 seconds. Also, wash your hair, scalp and eyebrows with an antibacterial shampoo.

**Omega-3s (fish oil)**

Some people find relief from their symptoms with omega-3 fatty acids, which may help the oil glands in the eyelids work better. Fatty fish like salmon or sardines contain omega-3s but you can also buy fish oil pills at the drugstore. Ask your doctor if they might help you.

**PREVENTION TIPS**

Many blepharitis cases aren’t preventable. Some risk factors for blepharitis, such as certain skin conditions, are beyond your control. Here are some steps you can take every day to help with the symptoms:

* Keep your hands, face and scalp clean.
* Try not to touch your itchy eyes or your face. Use a clean tissue if you must touch them.
* Remove all eye makeup before bedtime.
* Wipe away excess tears or eye drops with a clean tissue.
* Wear glasses instead of contact lenses until the condition clears.
* Use artificial tears if you have dry eyes and your provider agrees.
* Use anti-dandruff shampoo to wash your hair.
* Replace eye makeup — eyeliner, eye shadow, mascara — because they may have bacteria in the containers. You want to avoid reinfection.

**POSSIBLE COMPLICATIONS**

If you have blepharitis, you might also have:

* **Eyelash problems.** Blepharitis can cause your eyelashes to fall out, grow abnormally (misdirected eyelashes) or lose color.
* **Eyelid skin problems.** Scarring can develop on your eyelids from long-term blepharitis. Or the eyelid edges might turn inward or outward.
* **Excess tearing or dry eyes.** Abnormal oily secretions and other debris shed from the eyelids, such as flaking associated with dandruff, can build up in your tear film — the water, oil and mucus solution that forms tears.

Abnormal tear film interferes with keeping your eyelids moist. This can irritate your eyes and cause symptoms of dry eyes or excess tearing.

* **Stye.** A stye is an infection that develops near the base of the eyelashes. The result is a painful lump on the edge of your eyelid. A stye is usually most visible on the surface of the eyelid.
* **Chalazion.** A chalazion occurs when there's a blockage in one of the small oil glands at the margin of the eyelid, just behind the eyelashes. This blockage causes inflammation of the gland, which makes the eyelid swell and redden. This can clear up or turn into a hard, nontender bump.
* **Chronic pink eye.** Blepharitis can lead to recurrent bouts of pink eye (conjunctivitis).
* **Injury to the cornea.** Constant irritation from inflamed eyelids or misdirected eyelashes can cause a sore to develop on your cornea. Not having enough tears could increase your risk of a corneal infection

**When to see a doctor / red flag**

If you have blepharitis signs and symptoms that don't seem to improve despite good hygiene — regular cleaning and care of the affected area — make an appointment with your doctor.

**DIFFERENTIAL DIAGNOSIS**

* Allergic Contact Dermatitis
* Atopic Keratoconjunctivitis (AKC)
* Bacterial Conjunctivitis (Pink Eye)
* Bacterial Keratitis
* Basal Cell Carcinoma
* Chalazion
* Contact Lens Complications
* Dry Eye Disease (Keratoconjunctivitis Sicca)
* Epidemic Keratoconjunctivitis (EKC)
* Hordeolum
* Keratoconjunctivitis, Sicca
* Ocular Rosacea
* Preseptal Cellulitis
* Superior Limbic Keratoconjunctivitis (SLK)
* Trichiasis
* Viral Conjunctivitis (Pink Eye)

**EPIDEMIOLOGY**

Blepharitis is not specific to any particular group, affecting individuals of all ages, ethnicities, and genders. Estimates suggest that between 37% and 47% of patients treated by eye care specialists in the United States have blepharitis, making it one of the most common ocular disorders seen in clinical practice. Although blepharitis can affect people of all ages and ethnic backgrounds, its prevalence tends to increase with age, likely due to the impact of the normal aging process on meibomian gland function. The condition is most commonly seen in individuals aged 50 or older.

A major cross-sectional study conducted in Spain highlighted the prevalence of blepharitis, showing that symptomatic and asymptomatic MGD affected approximately 21.9% and 8.6% of the general population, respectively. A 10-year study conducted in South Korea (2004–2013) determined the overall incidence of blepharitis to be 1.1 per 100 person-years. The incidence increased over time and was higher in female patients. The prevalence for individuals aged 40 or older was 8.8%.

The epidemiology of blepharitis also appears to be influenced by gender. Some research suggests that women are more likely to be affected by certain subtypes of blepharitis, including staphylococcal blepharitis. For example, a single-center study found that women, with an average age of 42, comprised most patients with staphylococcal blepharitis. MGD and posterior blepharitis are more commonly observed in older populations, where age-related changes in glandular secretions and gland dysfunction are more prevalent. This condition, which links dermatological disorders with ocular manifestations, is also more frequently seen in populations with a higher incidence of rosacea

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**BLOCKED TEAR DUCT**

*ALTERNATIVE NAMES:* Blocked tear duct is also known as “dacryostenosis”; it can also be referred to as a “congenital lacrimal duct obstruction”.

**DEFINITION / DESCRIPTION**

A blocked tear duct is a condition that happens when something keeps tear fluid from draining out of your eyes properly. These blockages slow or stop the flow of tear fluid, causing it to back up in the tear duct system and into your affected eye. The medical term for this condition is “nasolacrimal duct obstruction.”

Your eyes need tear fluid to work properly. Your tear fluid lubricates the surface of your eye and helps your corneas absorb oxygen. Tear fluid also contains immune factors that protect against, or help your eyes recover from, infections.

You can think of your tear system like the gutters and downspouts on a house. The gutters channel water toward the downspout. But a blockage or clog in the downspout can make water back up and spill over the gutters’ edges.

The tear duct system is a series of openings and tubelike structures. The route that tear fluid follows to leave your eyes is:

* **The puncta**. Each eye has two puncta (the term for just one is “punctum”), one on your upper eyelid and another on the lower. They’re like the drain in a sink or bathtub, letting tear fluid flow out and into the canaliculi.
* **The canaliculi**. Each punctum drains into a canaliculus (the term for more than one is “canaliculi”). These canaliculi (pronounced “can-al-ICK-yew-lye”) merge and form a single tube before draining into the nasolacrimal ducts.
* **The nasolacrimal duct**s. The nasolacrimal (pronounced “nay-zo-LACK-rim-al”) ducts are the final tubelike areas that tear fluid travels through. At the bottom of each tear duct, there should be a valve.
* **The valve of Hasner**. This valve is just an opening that lets tear fluid drain into your nasal passages. Normally, it opens on its own before you’re born. But some babies have a valve that hasn’t opened yet. If it isn’t open, it’s called the membrane of Hasner.

Tear duct blockages are common in very young children, affecting between 6% and 20% of infants. They’re much less common in adults.

**CAUSES**

Tear duct blockages can happen for a few different reasons. Some are congenital, meaning you have them when you’re born. Others develop later in life.

**Congenital tear duct blockage**

Congenital tear duct obstruction usually happens because a child still has an unbroken membrane where the valve of Hasner should be. That means fluid can build up inside the lacrimal duct. Over time, the accumulated tear fluid can cause the duct to expand. It can also lead to an infection of the tear duct or other parts of the tear system.

**Other causes of tear duct blockage**

Tear duct blockages can also happen because of other conditions or be more likely to happen under certain circumstances. These are more likely to happen higher up in your tear duct system (or in parts that aren’t technically tear ducts, but they connect to the ducts).

Examples include:

* **Infections**. Chronic infections of your eyes or surrounding areas can contribute to scar tissue buildup. Sometimes, that scar tissue can cause narrowing and form a blockage.
* **Injuries**. Trauma to your face, eye or nose can cause swelling and tissue changes around your tear system. Swelling or tissue changes affecting any part of the system can cause or contribute to a blockage.
* **Narrow tear ducts (dacryostenosis)**. Sometimes, your tear ducts are narrow when you’re born, or they get narrower during your lifetime. Either can cause or contribute to a blockage.
* **Aging**. Your tear ducts and surrounding areas can change and narrow as you age.
* **Tumors or growths**. This includes benign (harmless) growths or cancerous tumors (but these are rare). Growths called mucoceles can also cause blockages in your tear ducts. These are small, mucus-filled pockets that can develop in the neighboring sinuses (the ethmoid sinuses on either side of your nose are right next to the tear system).
* **Tear stones (dacryoliths)**. These can form when tear fluid collects and hardens. If these are big enough, they can block a tear duct.

**RISK FACTORS**

Babies have the highest chance of developing a blocked tear duct. Blockages usually happen because the membrane at the bottom of the tear nasolacrimal duct doesn’t open and become the valve of Hasner.

Adults are more likely to develop a tear duct blockage if they have a history of:

* Chronic eye inflammation, such as uveitis.
* Glaucoma.
* Eye or sinus surgery.
* Previous cancer treatment, such as radiation therapy or chemotherapy.

**SIGNS / SYMPTOMS**

The symptoms of a blocked tear duct can include:

* Watery eyes (epiphora).
* Gooey or crusty buildup on your eyelids or in your eyelashes.
* Frequent rubbing of your eye or face around the blocked duct.
* Redness and swelling (from rubbing).
* Blurred vision.

These blockages can also make it easier to develop infections in the tear ducts. Infection symptoms can include:

* Swelling or redness of the tear duct or nearby tissues.
* Fever.
* Fussiness or irritability (especially in babies).
* Eye pain or sinus pressure.
* Redness or irritation of the eyeball, especially the sclera (the white part of your eye).

**DIAGNOSIS METHODS**

An eye care specialist or healthcare provider can usually diagnose a blocked tear duct based on your symptoms and by inspecting or feeling around your eye and tear duct. They’ll also ask you about your symptoms. If your baby might have a blocked tear duct, they can also check for this or do certain tests to look for a blockage.

One simple test they can do is called the “dye disappearance test.” To do it, a provider adds a drop of a special dye called fluorescein to your eye. Fluorescein glows under a blue light, so a provider can put a single drop of it into saline and then put the saline into your eye. After five minutes, they can check with a black light and see if any dye remains. If it’s still there, that can indicate you have a total or partial tear duct blockage.

Other tests are also possible, especially if your healthcare provider suspects that a blocked tear duct might be happening because of another condition or issue. The tests they recommend depend on your symptoms, circumstances and what condition they suspect could be the cause.

Because so many factors can play a role, it’s best to talk with them about the testing options. They can provide information that’s specific and relevant to your particular situation.

**TREATMENT OPTIONS**

A blocked tear duct is very treatable. But the treatments are different depending on the age of the person who has it and other factors. This condition can get better without treatment, especially in babies, but it’s better to try and treat it to avoid complications like infections.

Some treatments, like antibiotics, are possible no matter your age. These are most likely if you have an infection related to a blocked tear duct, and they can come in topical or eye drop forms. Other medications may also help. Your eye care specialist is the best person to tell you more about these options.

The main treatment for babies and infants with a blocked tear duct is a special massage technique that you can do at home. If your child or a child you care for has it, their pediatrician can show you exactly how to do this. This massage technique is usually all it takes to make the membrane pop open and become a valve.

If the blockage doesn’t get better with massage by the time a child has their first birthday, it’s more likely they’ll need a different treatment approach. Massaging also isn’t likely to help adults, so other treatments are usually necessary.

Some of these treatments may need to happen under sedation or anesthesia. Your (or your child’s) eye care specialist or provider can tell you if and why they recommend sedation or anesthesia.

**Dilation, probing and irrigation**

One of the simpler approaches to treating a blocked tear duct is a three-step technique called dilation, probing and irrigation. This technique can help babies if massage isn’t helpful. It’s also a likely first treatment for older children and adults. The steps of the technique work like so:

* **Dilation**. To do this first step, an eye care specialist will dilate (widen) the puncta and other tear system structures on the affected side. That makes room for what will happen in the next step.
* **Probing**. For this step, an eye care specialist will use the special tool(s) to probe (explore) the blocked part of the tear system. That helps them find the cause of the blockage and remove it, if possible.
* **Irrigation**. This final step involves pushing saline fluid through the tear duct to make sure it’s clear and wide enough.

**Balloon dilation**

In cases where simple dilation, probing and irrigation aren’t enough, a more advanced type of dilation may be necessary. When this is the case, your child’s eye care specialist will use a tool with a tiny balloon attachment on it. They’ll inflate the balloon slowly and gently in narrowed areas. Pushing like this from the inside can often widen the duct and resolve the issue.

**Stenting or intubation**

In some cases, balloon dilation may not be enough to widen a narrow tear duct permanently. In these cases, inserting a tube or stent might be the best approach. These devices provide a sturdy, hollow framework that can hold the duct open and still let tear fluid flow through.

**Surgical procedures**

In some cases, surgery may be the best choice to fix a blocked tear duct or reroute tear fluid around it. Two main surgeries are possible:

* **Dacryocystorhinostomy (DCR)**. For this procedure, a surgeon creates a bypass route for tear fluid to flow through. The new route will completely bypass the existing blockage.
* **Conjunctivodacryocystorhinostomy (CDCR)**. This procedure reconstructs your entire tear duct system.

The advantages, disadvantages and possible complications of the surgical procedures vary widely. The type of blockage you have, where it is and other factors can all play a role. Your surgeon or eye care specialist is the best source of information about what you can expect.

**PREVENTION TIPS**

Tear duct blockages happen for reasons that are unpredictable, outside your control or that you can’t see happening. That means there’s no way to prevent them.

There are a few things you can do to reduce the odds of an issue that could lead to a blockage, though. These mainly revolve around avoiding infections, including the following precautions:

* Avoid rubbing or excessively touching your eyes.
* Avoid sharing eye products, such as eye drops or cosmetics.
* Clean contact lenses according to your eye care provider’s instructions.
* Replace cosmetics — such as mascara, eyeliner or eyeshadow — every three to six months.
* Wash your hands frequently and thoroughly.

**OUTLOOK / PROGNOSIS**

Tear duct blockages usually have a positive outlook, depending on why they happen. Tear duct blockages aren’t dangerous on their own. But some of the causes can be. And blockages can lead to dangerous infections, so getting a blockage diagnosed and treated is important.

Congenital blockages have an excellent outlook. About 70% of children with these blockages get better by the time they’re 6 months old, and 90% get better by their first birthday. While blockages can go away spontaneously, the massage technique speeds up that process and reduces the chance of an infection or other issues. Congenital blockages also usually don’t cause problems later in life.

In adults, the outlook is generally good, depending on the cause. Most causes, especially injury-related causes, respond well to treatment, and many approaches can help. Your eye care specialist can tell you more about which treatments are most likely to help, the outcome you can expect, and what you can do to improve that outcome.

**Living With**

If you have (or a child you care for has) a tear duct blockage, an eye care specialist can guide you on treating and managing it. You shouldn’t try to self-diagnose or self-treat a tear duct blockage. If you think you have symptoms of a blockage, you should schedule an appointment with an eye care specialist and see them as soon as possible.

**When should I see my healthcare provider, or when should I seek care?**

If you have a tear duct blockage, your eye care specialist will give you treatment instructions and guidance. You can also ask them about the signs and symptoms that mean you need to call them or seek medical care more quickly.

The **differential diagnosis** is broad and includes conjunctivitis, corneal abrasion, uveitis hemangiomas, dermoid, nasal gliomas, and infantile glaucoma. The absence of associated signs and symptoms distinguishes congenital nasolacrimal duct obstruction (NLDO) from other causes of persistent tearing.

Most infants with congenital NLDO can be diagnosed and managed by the primary care practitioner. Referral to an ophthalmologist is warranted if the diagnosis is uncertain, particularly if there is any concern of glaucoma or signs or symptoms of dacryocystitis or dacryocystocele.

## **DIfferential Diagnoses**

* Adult Blepharitis
* Allergic Conjunctivitis
* Bacterial Conjunctivitis (Pink Eye)
* Corneal Foreign Body
* Dacryocystitis
* Distichiasis
* Dry Eye Disease (Keratoconjunctivitis Sicca)
* Emergency Care of Corneal Abrasion
* Entropion
* Intranasal drug abuse
* Preseptal Cellulitis
* Viral Conjunctivitis (Pink Eye)

**POSSIBLE COMPLICATIONS**

A blocked tear duct is just the kind of place where bacteria find it easy to grow, so bacterial infections or abscesses are the main possible complications of a blocked tear duct. The infections can also affect your sinuses, eyes and other nearby tissues. That’s why treating and keeping the infection from spreading is very important

## **Epidemiology**

### Frequency

United States

Nasolacrimal drainage obstruction is relatively common, but the exact frequency is not known.

International

Worldwide incidence is unknown.

### Mortality/Morbidity

Epiphora can be a nuisance. If untreated, nasolacrimal duct obstruction can cause significant problems.

### Race

No predilection to race has been established.

### Sex

PANDO is more prevalent in women. SALDO has no predilection to gender.

### Age

Previous studies have noted a high incidence of PANDO in individuals aged 50-70 years.

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**CATARACTS**

**ALTERNATIVE NAMES:** The name “cataract” encompasses a range of eye diseases; the known ones are: “Nuclear cataracts”, “Cortical cataracts”, “Posterior capsular cataracts”, “Congenital cataracts”, “Secondary cataracts”, “Traumatic cataracts”, and “Radiation cataracts”.

**DEFINITION / DESCRIPTION**

A Cataract is when there is a clouding of the eye's lens, leading to vision loss and blindness if untreated. Early cataracts may be managed with an adjustment in glasses prescription, but surgery may be necessary when vision is significantly impaired. It is one of the leading causes of blindness worldwide and a major cause of vision loss in the U.S.

A cataract is a clouding of the lens of the eye, which is typically clear. For people who have cataracts, seeing through cloudy lenses is like looking through a frosty or fogged-up window. Clouded vision caused by cataracts can make it more difficult to read, drive a car at night or see the expression on a friend's face.

Most cataracts develop slowly and don't disturb eyesight early on. But with time, cataracts will eventually affect vision.

At first, stronger lighting and eyeglasses can help deal with cataracts. But if impaired vision affects usual activities, cataract surgery might be needed. Fortunately, cataract surgery is generally a safe, effective procedure.

**Types of cataracts**

Cataract types include:

* **Cataracts affecting the center of the lens, called nuclear cataracts.** A nuclear cataract may at first cause objects far away to be blurry but objects up close to look clear. A nuclear cataract may even improve your reading vision for a short time. But with time, the lens slowly turns more yellow or brown and makes your vision worse. It may become difficult to tell colors apart.
* **Cataracts that affect the edges of the lens, called cortical cataracts.** A cortical cataract begins as white, wedge-shaped spots or streaks on the outer edge of the lens cortex. As the cataract slowly grows, the streaks spread to the center and affect light passing through the lens.
* **Cataracts that affect the back of the lens, called posterior subcapsular cataracts.** A posterior subcapsular cataract starts as a small spot that usually forms near the back of the lens, right in the path of light. A posterior subcapsular cataract often affects your reading vision. It also may reduce your vision in bright light and cause glare or halos around lights at night. These types of cataracts tend to grow faster than others.
* **Cataracts you're born with, called congenital cataracts.** Some people are born with cataracts or develop them during childhood. These cataracts may be passed down from parents. They also may be associated with an infection or trauma while in the womb.

These cataracts also may be due to certain conditions. These may include myotonic dystrophy, galactosemia, neurofibromatosis type 2 or rubella. Congenital cataracts don't always affect vision. If they do, they're usually removed soon after they're found.

**CAUSES**

Most cataracts develop when aging or injury changes the tissue that makes up the eye's lens. Proteins and fibers in the lens begin to break down. This causes vision to become hazy or cloudy.

Some disorders passed down from parents that cause other health problems can increase your risk of cataracts. Cataracts also can be caused by other eye conditions, past eye surgery or medical conditions such as diabetes. Long-term use of steroid medicines also may cause cataracts to develop.

**How a cataract forms**

A cataract is a cloudy lens. The lens sits behind the colored part of your eye, called the iris. The lens focuses light that passes into your eye. This produces clear, sharp images on the back part of the eye, called the retina.

As you age, the lenses in your eyes become less flexible, less clear and thicker. Aging and some medical conditions can cause proteins and fibers within the lenses to break down and clump together. This is what causes the clouding in the lenses.

As the cataract grows, the clouding becomes worse. A cataract scatters and blocks the light as it passes through the lens. This prevents a sharply defined image from reaching your retina. As a result, your vision becomes blurred.

Cataracts usually happen in both eyes, but not always at the same rate. The cataract in one eye may be worse than the other. This causes a difference in vision between eyes.

**RISK FACTORS**

Factors that increase your risk of cataracts include:

* Increasing age.
* Diabetes.
* Getting too much sunlight.
* Smoking.
* Obesity.
* Family history of cataracts.
* Previous eye injury or inflammation.
* Previous eye surgery.
* Prolonged use of corticosteroid medicines.
* Drinking excessive amounts of alcohol.

**SIGNS / SYMPTOMS**

Symptoms of cataracts include:

* Clouded, blurred or dim vision.
* Trouble seeing at night.
* Sensitivity to light and glare.
* Need for brighter light for reading and other activities.
* Seeing "halos" around lights.
* Frequent changes in eyeglass or contact lens prescription.
* Fading or yellowing of colors.
* Double vision in one eye.

At first, the cloudiness in your vision caused by a cataract may affect only a small part of the eye's lens. You may not notice any vision loss. As the cataract grows larger, it clouds more of your lens. More clouding changes the light passing through the lens. This may lead to symptoms you notice more.

**DIAGNOSIS METHODS**

To determine whether you have a cataract, your eye doctor will review your medical history and symptoms. They also will perform an eye exam. Your doctor may do several tests, including:

* **Vision test.** A vision test, also called a visual acuity test, uses an eye chart to measure how well you can read a series of letters. One eye is tested at a time, while the other eye is covered. A chart or a viewing device with letters that get smaller is used. With this, your eye doctor determines if you have 20/20 vision or if you have trouble seeing.
* **Eye structure exam.** An eye structure exam, also called a slit lamp, allows your eye doctor to see the structures at the front of your eye up close. It's called a slit lamp because it uses an intense line of light, a slit, to light up the structures in your eye. The slit allows your doctor to view these structures in small sections. This makes it easier to find anything that may be wrong.
* **Retinal exam.** A retinal exam looks at the back of your eyes, called the retina. To prepare for a retinal exam, your eye doctor puts drops in your eyes to open your pupils wide, called dilation. This makes it easier to see the retina. Using a slit lamp or a special device called an ophthalmoscope, your eye doctor can examine your lens for signs of a cataract.
* **Fluid pressure test.** This test, also called applanation tonometry, measures fluid pressure in your eye. There are multiple different devices available to do this.

**TREATMENT OPTIONS**

When your prescription glasses can't clear your vision, the only effective treatment for cataracts is surgery.

**When to consider cataract surgery**

Talk with your eye doctor about whether surgery is right for you. Most eye doctors suggest considering cataract surgery when your cataracts begin to affect your quality of life. This may include your ability to perform daily activities, such as reading or driving at night.

For most people, there is no rush to remove cataracts because they usually don't harm the eyes. But cataracts can worsen faster in people with certain conditions. These include diabetes, high blood pressure or obesity.

Waiting to do cataract surgery typically won't affect how well your vision recovers. Take time to consider the benefits and risks of cataract surgery with your doctor.

If you choose not to have cataract surgery now, your eye doctor may recommend periodic follow-up exams to see if your cataracts are getting worse. How often you'll see your eye doctor depends on your situation.

**What happens during cataract surgery**

Cataract surgery involves removing the clouded lens and replacing it with a clear artificial lens. The artificial lens, called an intraocular lens, is put in the same place as your natural lens. It remains a permanent part of your eye.

For some people, artificial lenses can't be used. In these situations, once the cataract is removed, vision may be corrected with eyeglasses or contact lenses.

Cataract surgery is typically done on an outpatient basis. This means you won't need to stay in a hospital after the surgery. During surgery, your eye doctor uses a medicine to numb the area around your eye. You usually stay awake during the procedure.

Cataract surgery is generally safe. However, it carries a risk of infection and bleeding. Cataract surgery also increases the risk of the retina being pulled out of place. This is called retinal detachment.

After the procedure, you may be sore for a few days. Healing usually happens within a few weeks.

If you need cataract surgery in both eyes, your doctor will schedule surgery to remove the cataract in the second eye after you've healed from the first surgery.

**Lifestyle and home remedies**

To deal with symptoms of cataracts until you decide to have surgery, try to:

* Make sure your prescription for your eyeglasses or contact lenses is up to date.
* Use a magnifying glass to read if you need more help reading.
* Improve the lighting in your home with more or brighter lamps.
* Wear sunglasses or a broad-brimmed hat to reduce glare during the day.
* Limit driving at night.

Self-care measures may help for a while, but your vision may get worse as the cataract grows. When vision loss starts to affect your everyday activities, consider cataract surgery.

**What you can do**

* **List any symptoms you're experiencing,** including any that may not seem related to the reason you scheduled the appointment.
* **Make a list of all medicines,** vitamins or supplements that you're taking.
* **Take a family member or friend along.** Sometimes it can be difficult to absorb all the information provided during an appointment. Someone who comes with you may remember something that you missed or forgot.
* **List questions to ask** your health care team.

**PREVENTION TIPS**

No studies have proved how to prevent or slow the growth of cataracts. But health care professionals think several strategies may be helpful, including:

* **Regular eye exams.** Eye exams can help detect cataracts and other eye problems at their earliest stages. Ask your health care team how often you should have an eye examination.
* **Do not smoke.** Ask a member of your health care team how to stop smoking. Medicines, counseling and other strategies are available to help you.
* **Manage other health problems.** Follow your treatment plan if you have diabetes or other medical conditions that can increase your risk of cataracts.
* **Choose a healthy diet that includes plenty of fruits and vegetables.** Adding fruits and vegetables to your diet ensures that you're getting many vitamins and nutrients. Fruits and vegetables have antioxidants. Antioxidants help maintain the health of your eyes.

Studies haven't proved that antioxidants in pill form can prevent cataracts. But a large population study recently showed that a healthy diet rich in vitamins and minerals reduced the risk of developing cataracts. Fruits and vegetables have many proven health benefits. Eating them is a safe way to get enough minerals and vitamins in your diet.

* **Wear sunglasses.** Ultraviolet light from the sun may cause cataracts. Wear sunglasses that block ultraviolet B rays when you're outdoors.
* **Reduce alcohol use.** Drinking too much alcohol can increase the risk of cataracts.

**Prognosis**

An ophthalmologist or optometrist is the best person to ask about your outlook. They’ll examine your eyes and the severity of any cataracts you have.

Early on, you might not need surgery. The changes to your vision may be mild. But cataracts can progress over time, causing more noticeable symptoms. If your symptoms start to interfere with your daily life, your provider may recommend surgery to help you safely go about your usual tasks.

**WHEN TO SEE A DOCTOR**

Make an appointment for an eye exam if you notice any changes in your vision. If you develop sudden vision changes, such as double vision or flashes of light, sudden eye pain, or a sudden headache, see a member of your health care team right away.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis of cataracts includes many disorders, such as:

* Glaucoma
* Refractive errors
* Macular degeneration
* Diabetic retinopathy
* Corneal dystrophies and degenerations
* Optic atrophy
* Retinitis pigmentosa

**Recent guidelines or updates**

First visit (24-36 hours postoperative). At this visit, the patient's visual acuity, both unaided and with a pinhole, is measured. Keratometry and retinoscopy may be performed. The IOP is tested and the anterior segment is examined to ensure IOL centration and intactness of the structures. The following structures should be evaluated with the slit lamp: conjunctiva, cornea, anterior chamber, IOL, capsule, and wound. Funduscopic examination is performed when there are symptoms of very poor vision or retinal disease. At the conclusion of the first postoperative visit, the patient is counseled regarding follow up care (e.g., instructed in how to use the antibiotic and/or steroid drops, advised concerning the level of physical activity permitted, and warned of symptoms that require emergency care) and the second postoperative visit is scheduled.

• Second visit (7-14 days postoperative). Visual acuity, both unaided and with a pinhole, should be measured . Tonometry and slit lamp examination should be performed as outlined for the first visit. Dilated fundus examination should be performed when indicated by signs or symptoms of retinal disease. At the conclusion of the second visit, the patient should be informed of his or her progress and instructed to continue or taper the antibiotic and/or steroid drops. If only an antibiotic drop was prescribed, it may be discontinued at this point. In the case of a steroid-only drop or an antibiotic/steroid combination drop, tapering may begin if the eye is quiet. This is generally accomplished by decreasing the dosage frequency by 1 less daily drop each week.73 The patient should again be advised regarding the level of physical activity permitted and warned of symptoms requiring emergency care. An appointment should be scheduled for the third visit.

Third visit (3-4 weeks postoperative). The examination and instructions to the patient are the same as for the second visit. A refraction may be performed, and spectacles may be prescribed if the eye appears quiet and stable. An appointment should be scheduled for the fourth visit, if needed.

• Fourth visit (6-8 weeks postoperative). Examination and instructions to the patient are the same as for the second visit. A dilated fundus examination is recommended at the final postoperative visit if one has not been done earlier. If the patient's eye is quiet, any topical medications still in use can be discontinued. A refraction should be performed, and the patient may be given the final postoperative prescription, if it was not given at the previous visit. Spectacle or contact lens correction is usually needed after cataract surgery with IOL implantation to correct any residual refractive error or pseudophakic presbyopia. Residual refractive error may be due to planned or unexpected undercorrection or overcorrection by the IOL power and/or due to pre-existing corneal astigmatism or induced corneal astigmatism caused by suturing of the incision. A near vision prescription may also be needed to compensate for the loss of focusing power of the eye due to removal of the lens. Some ophthalmic surgeons prefer to set a goal of low myopia (less than 1 diopter [D]) rather than emmetropia for postoperative refractive error. This may allow the patient to function at many intermediate and near visual tasks without a lens prescription. The "optimal" postoperative refraction for a particular patient may be based on the refractive status of the other eye, whether cataract surgery is planned or anticipated for both eyes, and the specific visual status and needs of the patient. There are several special considerations for prescribing glasses for patients who have undergone cataract extraction with IOL implants. For patients who have had surgery in one eye only, more than 1.50 D of anisometropia may result in asthenopia related to correction with different spectacle lens powers. The option of the use of a contact lens on the fellow eye should be discussed with the patient. In The Care Process Cataract patients with presbyopia undergoing cataract extraction in one eye only, the fellow eye typically has a near add power of approximately 2.50 D, which allows a normal correction for the operated eye of approximately 2.50 D, bringing the two eyes into balance at near. However, for younger patients who have some residual accommodative function, a monocular add for the operated eye is required to bring it into focus with the fellow eye when reading. Ultraviolet protection is provided in most implanted IOLs. This protection is needed in the replacement lens because the natural lens normally absorbs UV radiation and shields the retina from exposure. For patients who have IOLs without UV absorption, the benefits of UV protection (e.g., reducing the risk of age-related macular degeneration) should be discussed. UV protection can also be provided in spectacles and contact lenses. For patients who have anisometropia of 3 D or less, vision can usually be satisfactorily corrected with spectacle lenses. In patients with greater differences between lens powers, and even in some patients whose lens powers differ by as little as 1 D, symptoms of diplopia and asthenopia (particularly at near), headache, or photophobia may occur. Aniseikonia also needs to be considered in the spectacle management of these patients. If spectacle management is not satisfactory, a contact lens, IOL exchange, or refractive surgery may be considered to balance the refractive correction between the two eyes. Postoperative astigmatism may be influenced by size or location of the incision, suturing method, or suture tension. When astigmatism is caused by tight suturing, sutures may be cut after approximately 4-6 weeks. Often this can reduce astigmatism, which typically has the steepest corneal meridian directly aligned with the orientation of the tightest suture. When induced corneal steepening in the vertical meridian is associated with tight sutures, suture-cutting should be considered before glasses are prescribed to reduce the astigmatism in the spectacle correction.

**EPIDEMIOLOGY**

Many studies in 2010 reveal that cataracts are most common in the White American race, where prevalence ranges from 17 to 18% per 100 people. Blacks were the second-highest affected by cataracts, with a 13% prevalence rate, followed by Hispanics, with a prevalence rate of almost 12%.

**Age:** Onset is gradual and progressive, commonly in the older age group, typically in the fifth and sixth decade, though cases have been reported in children and the elderly as well.

**Sex:** Recent studies reveal that the disease is more common in women than men, with a male-to-female ratio of 1 to approximately 1.3

Prevalence and Incidence Studies on the prevalence of cataract have focused on different sample populations:

• The National Health and Nutritional Examination Survey (NHANES) studied both genders and all races, sampled from a broad range of communities.

• The Watermen Eye Study included men only from a selected region.

• The Framingham Eye Study included both genders in a small community.

• The Beaver Dam Eye Study included both genders in a rural community

The NHANES study showed a progressive increase in lens opacities with age. Approximately 12 percent of participants of ages 45-54, 27 percent of those ages 55-64, and 58 percent of those ages 65-74 had lens opacities. Of the 65-74 year age group, 28.5 percent had lens opacities with associated vision decrease.24 The Watermen Eye Study examined lens opacities for fishermen in age ranges from 30 to 94 years and found a progressive increase in lens opacities with age. Cataract was present in approximately 1.8 percent of men under the age of 35 years. Lens opacities causing vision loss were found in approximately 5 percent of the age 55-64 group, 25 percent of the age 65-74 group, and 59 percent of the 75-84 age group.25 The Framingham Eye Study showed the prevalence of cataracts without vision loss ranged from 41.7 percent in persons ages 55-64 to 91.1 percent in persons ages 75-84. The prevalence of lens opacity with decreased vision was 4.5 and 45.9 percent, respectively, for the same age groups.26 The Beaver Dam Eye Study evaluated the prevalence of cataract in adults between the ages of 43 and 84 years. Overall, 17.3 percent had NS more severe than level 3 in a five-step scale of severity. The investigators found cortical opacities in 16.3 percent of this population, PSC in 6 percent. Women were more commonly affected than men.

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**CHARLES BONNET SYNDROME**

**DESCRIPTION / DEFINITION**

Charles Bonnet syndrome (pronounced “bo NAY”) is a condition that happens when a person with low vision has visual hallucinations.

The eye’s retina facilitates the conversion of light into a visual message for the brain. When you’re not able to do this, your vision gets worse and you become more likely to develop Charles Bonnet syndrome.

The hallucinations can be simple, like bright-colored patterns that show up over everything you see. The medical term for this is unformed hallucinations.

You can also see more complicated things, like animals, buildings or people that don’t exist. The medical term for this is formed from hallucinations.

Although scientists aren’t quite sure of the pathophysiology behind Charles Bonnet syndrome, they think that the hallucinations happen because lower vision reduces the number of images your brain gets. Your brain wants more images, so it uses memories to recall things that it’s seen or simply makes things up.

This can be worrisome and confusing if you don’t know what’s happening.

Charles Bonnet syndrome can affect people with conditions that cause vision loss, such as age-related macular degeneration (ARMD). One study indicated that more than 12% of people with ARMD will develop Charles Bonnet syndrome.

An estimated 1 in 2 people with severely impaired vision may develop hallucinations. Charles Bonnet syndrome can happen in anyone, but it’s more frequent in patients 80 years or older.

In addition to AMRD, other conditions related to developing Charles Bonnet syndrome include:

* Stroke.
* Cataracts.
* Glaucoma.
* Retinitis pigmentosa.
* Diabetes-related retinopathy.
* Optic neuritis.
* Giant cell arteritis.
* Eye removal.

**CAUSES**

The cause of Charles Bonnet syndrome is related to loss of vision. A main theory is that your brain wants images and isn’t getting enough, so it creates them.

Some studies have indicated that what you see suppresses some nerve activity in your brain. When your eyes aren’t supplying that type of suppression, nerve activity in the brain appears as visual hallucinations.

**RISK FACTORS**

Risk factors for Charles Bonnet syndrome include:

* Being 80 years old or older.
* Having low vision.
* Being alone/socially isolated.
* Being in a dark environment.

**SIGNS / SYMPTOMS**

The key symptom of Charles Bonnet syndrome is seeing hallucinations. These hallucinations may appear as:

* Patterns made up of lines or shapes that repeat. These images might look like bricks or nets.
* Imaginary animals like dragons or unicorns.
* Outdoor scenes, like waterfalls, trees or mountains.
* People’s faces, people, animals or bugs.
* People dressed as if they were living in the past.

The images may:

* Move or stay in one place.
* Be in color or be in black and white.
* Be true to size or be smaller than normal.
* Be pleasant or be scary.
* Be familiar to you or not familiar to you.
* Be the same each time they happen or change each time they happen.

These visions usually happen without noise. They can continue for various lengths of time from seconds to minutes to hours.

Most people with Charles Bonnet syndrome know that what they’re seeing isn’t real.

People with Charles Bonnet syndrome can’t control the hallucinations.

**DIAGNOSIS METHODS**

Your healthcare provider or eye care specialist will take your medical history, ask questions about your symptoms and do thorough physical and eye examinations.

It’s possible to have hallucinations when you’re taking certain medications. It’s also possible to have hallucinations if you’re withdrawing from using drugs or alcohol. You can also have hallucinations after cataract surgery.

Your providers will want to eliminate any other conditions that could possibly cause hallucinations, such as Alzheimer’s disease, Parkinson’s disease, Lewy body dementia and schizophrenia.

Your providers may recommend neurological tests to rule out these other diseases.

**TREATMENT OPTIONS**

**Management and Treatment**

There’s no cure for Charles Bonnet syndrome. Providers have tried medications but they didn’t work. Symptoms often improve with time, possibly because your brain becomes used to receiving fewer images to process. However, it may take years for the hallucinations to stop.

If you have Charles Bonnet syndrome, you may be able to manage symptoms better with the following tips.

**Managing Charles Bonnet syndrome with eye movements**

You may want to blink your eyes or close your eyes. You might want to look away from the hallucination or stare at it. You can also try moving your eyes without moving your head, looking from side to side or up and down.

**Managing Charles Bonnet syndrome with improved lighting**

You may find it easier to deal with Charles Bonnet syndrome if you have brighter light, especially in the evenings.

**Managing Charles Bonnet syndrome by being less isolated**

It may help you to spend less time alone and spend more time with people. The people you spend time with should be people you can talk to about your life, including the images you’re seeing. You may want to find a support group to improve your connections.

**Managing Charles Bonnet syndrome with rest and relaxation**

Some people have found that their symptoms get worse when they’re under stress or don’t get enough rest. Try to get enough sleep and to develop other ways to relax and to relieve stress. These could include practices like meditation, deep breathing and learning to relax your muscles in a progressive way.

In addition to these suggestions, your provider may suggest anti-anxiety medications or antidepressants to help you deal with your emotions and uncertainties.

**PREVENTION TIPS**

Since researchers don’t really know what causes Charles Bonnet syndrome, there’s no concrete way to prevent it. You may be able to reduce your risk by taking certain steps to preserve your vision. For instance, if you have diabetes, you can do your best to manage your blood sugar levels.

It’s very important for everyone to have regular eye exams. This is especially true as people age, since many of the causes of vision loss happen as people get older.

It’s important to have a good relationship with your ophthalmologist or other eye care provider.

**OUTLOOK / PROGNOSIS**

There’s no cure for Charles Bonnet syndrome. It may disappear over time, but it may take one to two years to go away.

You may find it difficult to do some of your daily tasks or walk in places that you don’t know well if you’re seeing things that aren’t there. It’s important to tell your healthcare provider about it.

**Living With**

**What questions should I ask my healthcare team about Charles Bonnet syndrome?**

You may have many questions for your providers about Charles Bonnet syndrome. Some of them may include:

* Are there certain tests you would recommend to determine if I actually do have Charles Bonnet syndrome?
* Can you recommend a support group for me?
* Would I benefit from being in a clinical trial?
* How can I deal with the condition that’s causing my vision loss?
* Can you suggest ways for me to deal with Charles Bonnet syndrome?

**What is the difference between Charles Bonnet syndrome and Anton syndrome?**

If you have Charles Bonnet syndrome, you see hallucinations and you know that they’re not real. If you have Anton syndrome, you can’t see but claim that you can. You tell people that you’re seeing things that you really aren’t seeing. You deny your blindness, which has often occurred after a cerebrovascular injury.

**DIFFERENTIAL DIAGNOSIS**

Essential considerations for differential diagnosis pertain to etiologies involving visual hallucinations. These include

* Narcolepsy
* Peduncular hallucinosis
* Levodopa-induced hallucinations
* Hypnagogic hallucinations
* Migraine coma
* Schizophrenia
* Epileptic seizures
* Dementia
* Migraine aura
* Neurodegenerative - Parkinson, Alzheimer, and Lewy body dementia
* Metabolic encephalopathy - drugs, alcohol withdrawal, or delirium

CBS is differentiated from all these conditions by the simultaneous presence of visual deficits and the absence of neurological deficits. Furthermore, CBS is differentiated by the lack of auditory or sensory-associated hallucinations, which can be seen in many of the above differentials.

**EPIDEMIOLOGY**

The incidence and prevalence of CBS are still under investigation and require further research. However, various studies and surveys indicate that the condition is not as uncommon as one might think. CBS prevalence has been found to differ widely depending on different sample sizes, with estimates ranging from 1% to nearly 10%.

One large study aimed at investigating CBS found the overall prevalence to be 0.5% (5/1000), with 0.8% (1/120) of their sample being in those with low visual acuity, 0.6% (2/346) being in the elderly, and 0.8% (1/120) being in both the elderly and those with low visual acuity.

Further highlighting the predominance in those with a higher degree of visual impairment or even more in those with complete vision loss, a study comparing visual hallucinations in people with macular disease versus people with glaucoma involved in a visual rehabilitation found the prevalence of CBS to be 39% and 20% respectively.

Regarding age, CBS can affect all ages; however, it tends to be more prevalent in older individuals. Data on CBS shows the mean age of the condition to be 70 to 85 years old, the expected range where pathological processes leading to visual impairment or vision loss naturally occur.

However, literature has suggested that CBS is vastly under-reported, mainly due to patients fearing being diagnosed with a psychiatric illness. For instance, one study found that a significant number of patients are unaware of the disease, with only 12% of patients in a retinal clinic knowing of CBS. In contrast, another study found that visual hallucinations were reported in only 11 to 15% of older individuals with impaired vision.

CBS is more common in older patients. Previous reports have revealed the mean age of incidence ranging from 74 to 84 years. CBS has also been reported in children having a high incidence of rapid visual loss. The incidence of profound visual loss is also reported more commonly in the elderly. De Morsier noted a male preponderance; some studies showed a female preponderance, while some showed no sex bias.

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**CENTRAL SEROUS RETINOPATHY**

*ALTERNATIVE NAMES:* Central serous retinopathy is also known as “Central serous chorioretinopathy”, or “Central serous choroidopathy”.

**DEFINITION / DESCRIPTION**

Central serous retinopathy is a medical condition that occurs when fluid builds up behind the retina in your eye. The fluid comes from a layer of blood vessels beneath your retina called the choroid. This fluid can cause your retina to detach, leading to vision loss or other vision problems.

Your retina is a layer of tissue behind each eye. It senses light and translates it into images your brain can understand.

Central serous retinopathy affects about 10 out of every 100,000 people, depending on your sex and other risk factors.

**CAUSES**

**Central serous chorioretinopathy causes**

Scientists don’t fully understand what causes central serous retinopathy. Stress appears to play a key role. Stress makes your body release a hormone called cortisol, which can cause inflammation and fluid leakage. People with high stress levels and reduced coping skills are at a higher risk for central serous retinopathy.

The condition is also associated with the use of medications containing corticosteroids, which treat inflammation.

**RISK FACTORS**

Central serous retinopathy can affect anyone, but it’s more common in:

* Males between the ages of 30 and 50
* People with myopia (nearsightedness)
* Those taking certain medications, especially corticosteroids

Other risk factors include:

* Autoimmune diseases, like lupus and rheumatoid arthritis
* Heart disease or high blood pressure
* Infection with the bacterium *Helicobacter pylori*
* Kidney disease, like glomerulonephritis
* Pregnancy
* Sleep problems, like sleep apnea and insomnia
* Use of certain medications, like those that treat nasal congestion and erectile dysfunction

**SIGNS / SYMPTOMS**

Central serous retinopathy can affect one eye or both at the same time. Central serous chorioretinopathy symptoms may include:

* Blurry vision, like a smudge in the center of your sight
* Dark spot in the center of your vision
* Darker or dim vision
* White items look dull or somewhat brown
* Objects seem smaller or further away than they are
* Straight lines look crooked or bent

But central serous chorioretinopathy doesn’t always cause symptoms. A person can have the condition but not have vision problems.

**DIAGNOSIS METHODS**

If you have changes to your vision, seek medical attention from your primary care provider or an ophthalmologist (eye specialist). They’ll talk to you about your symptoms and perform an eye exam. They may also order certain retina tests, including:

* **Fluorescein angiography (IVFA)**.A healthcare provider injects a dye into a vein in your arm that spreads to your retina. Then, they take pictures with a special camera to identify leaks.
* **Optical coherence tomography (OCT)**. A scan that provides 3D pictures of your retina so your provider can measure retinal thickness, identify swelling and detect serous retinal detachment.

**TREATMENT OPTIONS**

**Management and Treatment**

Many cases of central serous retinopathy go away on their own over a few weeks or months. Your healthcare provider may recommend monitoring, or a “watch and wait” approach. During the monitoring period, they’ll repeat tests to ensure the fluid is draining.

Your provider may also ask you to stop taking medications that contribute to the eye condition. They’ll counsel you to reduce your stress levels.

If the fluid doesn’t drain on its own in a few months, your provider may recommend central serous chorioretinopathy treatment, including:

* **Medications**.Some medications, like anti-vascular endothelial growth factors, can prevent new blood vessels from growing in your eyes. Certain diuretics can help reduce fluid.
* **Photodynamic therapy (PDT)**. A healthcare provider injects a drug called verteporfin into your arm that travels to your eye. Then, they use a cold laser to close the leak.
* **Other laser treatments**.After giving you numbing medications, your provider may use thermal laser treatment (a heated laser) to seal leaks. Micropulse laser uses smaller, shorter laser pulses.

**How do I take care of myself with central serous retinopathy?**

Some lifestyle changes can help you manage central serous retinopathy:

* Get at least seven hours of sleep every night
* Limit caffeine, alcohol and corticosteroids
* Manage and reduce stress with exercise, meditation, time with loved ones or counseling

If you’re experiencing vision loss, some coping strategies include:

* Joining a support group to meet people who understand
* Talking to loved ones or a counselor about your feelings and ways to cope
* Using aids to help you complete daily tasks safely
* Prevention tips

**OUTLOOK / PROGNOSIS**

Many cases of central serous retinopathy go away on their own as fluid naturally drains. But treatment may be necessary if the condition continues for several months.

After the condition resolves, vision generally improves on its own, often returning to normal. But sometimes, damage can be permanent, with vision changes that don’t improve.

The condition can happen again, even after successful treatment, in the same eye or your other eye. For this reason, you may need regular follow-up appointments with an ophthalmologist.

**WHEN TO SEE A DOCTOR**

If you experience any changes to your vision, talk to your primary care provider or an ophthalmologist.

Even though central serous chorioretinopathy can go away on its own, it can get worse and cause permanent vision changes or loss. And sometimes, vision problems are a sign of underlying disease.

**Differential diagnosis (how it’s distinguished from other illnesses)**

Other diseases of choroid and retina can closely mimic CSCR. They are ARMD (age-related macular degeneration), IPCV, and optic disc pit associated maculopathy. ARMD is seen in patients above 50 years. Chronic CSCR can develop secondary CNVM on follow up or after laser photocoagulation.

OCT angiography is helpful in delineating the neovascular complex of ARMD. EDI OCT shows thicker choroid in CSCR and thin to normal choroid in ARMD. IPCV produces serous macular detachment and RPE changes similar to CSCR. The polyp and branching vascular network are characteristic findings on ICGA. OCT typically shows serosanguinous or notched PED.

Optic disc pit causes serous macular detachment at the macula. On careful slit-lamp biomicroscopy, focal excavation can be seen on the temporal part of the optic disc. Retinoschisis is commonly observed in the optic disc pit, while these changes are infrequently seen only in chronic CSCR. The sub-internal limiting membrane cavity after the resolution of sub-internal limiting membrane hemorrhage (including after Valsalva retinopathy) may simulate CSCR on cursory examination.

**Epidemiology data**

CSCR is the fourth most common retinal disorder threatening the vision.Men are commonly affected. The male-female ratio was found to be 6:1 in a population-based study. The mean age group is 39-51 years. When females are affected, the age is usually higher than males. It is generally unilateral. However, bilateral involvement may be in up to 40% of cases. However, the majority of cases have pigment epithelial detachment (PED) in the fellow eye also. Bilateral changes in the choroid are usually noted on optical coherence tomography (OCT) and indocyanine green angiography (ICGA).Kitzmann et al. found the incidence of CSCR to be 9.9 cases per 100,000 population

**REFERENCES**

<https://my.clevelandclinic.org/health/diseases/24335-central-serous-retinopathy>

[Central Serous Chorioretinopathy - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK558973/#article-96027.s9)

**CHALAZION**

ALTERNATIVE NAMES: Chalazion is also known as a “meibomian cyst”, “meibomian cyst”, “lipogranuloma”, or “conjunctival granuloma”.

**DEFINITION / DESCRIPTION**

Chalazion is a condition where the oil glands in the eyelid become blocked causing a lump, which causes a red bump on your eyelid. It’s sometimes called an eyelid cyst or a meibomian cyst. It slowly forms when an oil gland (called a meibomian gland) becomes blocked. At first, the chalazion may be painful, but after a little time, it usually doesn’t hurt. A chalazion typically forms on the underside of your upper eyelid but may occasionally form on your lower eyelid.

Ordinarily, chalazia’s (the plural of chalazion) develop in adults between the ages of 30 and 50. They’re not common in children, but they can happen.

#### Chalazion vs. stye — what’s the difference?

It can be hard to tell the difference between a chalazion and a stye. A chalazion can form because of a stye, but they’re different conditions. While they both form due to blocked oil glands, styes are bacterial infections that cause the gland to swell. Styes appear at your eyelid’s edge, whereas chalazia’s appear farther back on your eyelid. In addition, styes can be painful, but chalazia generally aren’t painful.

**CAUSES**

A chalazion develops when something blocks a small oil gland in your eyelid. These glands (meibomian glands) help keep your eyes moist. A blocked gland begins retaining oil and swells. Eventually, the fluid will drain, causing irritation to your surrounding eyelid skin. This irritation can lead to a hard lump filled with the oil and fluid on your eyelid.

**Are chalazions contagious?**

No, chalazia aren’t contagious. Bacterial infections don’t cause them, so bacteria can’t spread from person to person.

**RISK FACTORS**

Chalazia are very common, and anyone can get them. But you may be more likely to get a chalazion if you:

* Have had a chalazion before.
* Have chronic blepharitis (eyelid inflammation).
* Have certain skin conditions, like dandruff (seborrheic dermatitis) or rosacea.
* Have dry skin.
* Are experiencing hormonal changes.

**SIGNS AND SYMPTOMS**

If you have a chalazion, you’ll notice a bump on your eyelid. It usually affects your upper lid. When a chalazion first develops, it may be painful, but the pain will quickly go away. As the chalazion grows, it may become red, swollen and tender.

Other chalazion symptoms may include:

* Mild irritation, causing your eye to water.
* Blurred vision from a larger chalazion pushing on your eyeball.
* Entirely swollen eyelids.

**DIAGNOSIS METHOD**

You’ll usually see an eye care specialist when you have a chalazion. You might see an optometrist or an ophthalmologist. These healthcare providers can examine the chalazion and offer treatment options.

**When you see an eye care specialist, you can expect:**

* Health history: Your provider will go over your complete health history. This information can help your provider find underlying issues that could be contributing to the formation of a chalazion.
* External eye exam: Your provider will perform a complete eye exam, looking at your eye, eyelid, eyelashes and skin texture.
* Thorough eyelid exam: Your provider will shine a bright light and use magnification to look at the base of your eyelashes. They’ll also check the oil glands’ openings on the underside of your eyelid.

**TREATMENT OPTIONS**

In most cases, you can treat a chalazion at home. Most chalazion go away in a month or less.

First, never push on a chalazion or try to pop it. You can cause inflammation and infection. Instead, for chalazion self-care, try:

* Warm compresses: Wet a clean washcloth with warm water. Hold it on the affected eye for 15 minutes. Do this at least three times a day to help the blocked oil gland open up.
* Good hygiene: Don’t wear eye makeup while you have a chalazion. After the chalazion drains, keep the area clean. Follow good eye health practices, and avoid touching your eyes.

If the chalazion doesn’t go away, you should seek help from an eye care specialist. Chalazion treatment in your provider’s office may include draining the fluid through a small cut (incision). You may also need an injection of steroids to reduce swelling and inflammation.

**PREVENTION TIPS**

Some chalazia form because of styes. You may be able to avoid getting a chalazion if you can avoid getting a stye. You can avoid getting styes by following good hygiene. Some essential elements of good hygiene include:

* Hand washing: Wash your hands thoroughly and often. Before you touch your eyes, make sure to wash your hands.
* Contact lens care: Wash your hands before removing contact lenses. Make sure to thoroughly clean your contacts with a disinfectant and lens cleaning solution. Always throw daily and limited-time contacts away on schedule.
* Face washing: Wash your face daily to remove dirt and makeup before going to bed. Your healthcare provider may recommend cleaning your eyelids with a special scrub or baby shampoo, especially if you’re prone to blepharitis.
* Makeup hygiene: Throw away all of your old or expired makeup. Be sure to replace mascara and eye shadow every two to three months. Also, never share or use another person’s makeup.

**OUTLOOK / PROGNOSIS**

With proper home management, a chalazion should heal in a week. If left untreated, it can take four to six weeks for the chalazion to heal. Some can persist for many months.

**Will I get more chalazia?**

If you have one chalazion, you may get another. Always practice good hygiene to help prevent future chalazia.

**When should I see my healthcare provider?**

If you have a chalazion that doesn’t go away with home treatment, see an eye care specialist. They’ll be able to examine your eye and offer additional treatment options. You should also see your healthcare provider if you have recurring chalazia (eyelid bumps that come back).

**POSSIBLE COMPLICATIONS**

Larger chalazion can press on the surface of your eye (cornea), which can lead to blurred and decreased vision.

**DIFFERENTIAL DIAGNOSIS**

* Acute Complications of Sarcoidosis
* Adult Blepharitis
* Ptosis (Blepharoptosis) in Adults
* Allergic Contact Dermatitis
* Atopic Dermatitis in Emergency Medicine
* Bacterial Conjunctivitis (Pink Eye)
* Basal Cell Carcinoma
* Capillary Hemangioma
* Orbital Cavernous Hemangioma
* Congenital Anomalies of the Nasolacrimal Duct
* Conjunctival Melanoma
* Contact Lens Complications
* Dacryoadenitis
* Dacryocystitis
* Ocular Demodicosis (Demodex Infestation)
* Dermatochalasis
* Dermatologic Manifestations of Kaposi Sarcoma
* Distichiasis
* Eyelid myxoma
* [Eyelid Papilloma](https://emedicine.medscape.com/article/1211855-overview)
* Floppy Eyelid Syndrome
* Herpes Simplex Virus (HSV) in Emergency Medicine
* Herpes Zoster
* Hordeolum
* Sturge-Weber Syndrome Imaging and Diagnosis
* Juvenile Xanthogranuloma
* Lacrimal Gland Tumors
* Leishmaniasis
* Molluscum Contagiosum
* Nasolacrimal Duct Obstruction and Epiphora
* Ocular Manifestations of HIV Infection
* Ophthalmologic Manifestations of Neurofibromatosis Type 1 (NF-1)
* Orbital Cellulitis
* Orbital Dermoid
* Orbital Tumors
* Pediatric Actinomycosis
* Pediatric Tuberculosis
* Pigmented Lesions of the Eyelid
* Preseptal

**EPIDEMIOLOGY**

**United States and international statistics**

Chalazia are common, but their exact incidence and prevalence in the United States are not known. Data about the worldwide prevalence or incidence of chalazia also are unavailable.

**Age-related demographics**

Although chalazion occurS in all age groups, they are more common in adults (especially those aged 30-50 years) than in children, presumably because androgenic hormones increase sebum viscosity. ] Hormonal influences on sebaceous secretion and viscosity may explain clustering at the time of puberty and during pregnancy; however, the large number of patients without evidence of hormonal alteration suggests other mechanisms also apply. Chalazia are uncommon at the extremes of age, but pediatric cases may be encountered.

Recurrent chalazion, particularly in elderly patients, should prompt the practitioner to consider conditions that may masquerade as a chalazion (eg, sebaceous carcinoma, squamous cell carcinoma, microcystic adnexal carcinoma, tuberculosis). Recurrent chalazion in a child or young adult should prompt an evaluation for viral conjunctivitis and hyperimmunoglobulinemia E (hyper-IgE) syndrome (Job syndrome).

**Sex- and race-related demographics**

Chalazia appear to affect males and females equally, but as noted, precise information about prevalence and incidence is not available. Contrary to popular opinion, research has not shown that the use of eyelid cosmetic products either causes or aggravates the condition.

*REFERENCES:*

[**Chalazion Differential Diagnoses**](https://emedicine.medscape.com/article/1212709-differential?form=fpf)

[**Chalazion: Symptoms, Causes, Prevention & Treatments**](https://my.clevelandclinic.org/health/diseases/17657-chalazion)

**CHOROIDEREMIA**

*ALTERNATIVE NAMES:* Choroideremia is also known as CHM, and sometimes referred to as “Tape Choroidal dystrophy”.

**DEFINITION / DESCRIPTION**

Choroideremia (pronounced “kuh-roy-der-ee-me-ah”) is a rare inherited disease that affects your eyes. It causes vision to get worse and then leads to blindness. Severe disease mostly affects men.

Choroideremia is similar in some ways to retinitis pigmentosa, another condition that causes your retina to degrade. In fact, the two diseases have similar symptoms and sometimes have similar test results. Getting a genetic test is the best way to tell the difference between the two diseases.

In your eye, your retina has photoreceptor cells that capture light and turn it into electrical signals. Those signals travel along your optic nerve to your brain where they’re turned into images. The choroid is a layer between your retina and sclera, which is the white protective covering of your eye.

The choroid has blood vessels that feed your retina. Damage to your choroidal blood vessels leads to damage to your retina and peripheral sight.

Choroideremia affects an estimated 1 in 50,000 to 1 in 100,000 people

**CAUSES**

Choroideremia is an X-linked genetic disorder. The gene that causes choroideremia is located on your X chromosome. Females have two X chromosomes, while males have one.

Because there’s only one X chromosome with the faulty gene related to choroideremia, females don’t normally have the same vision problems that males with the faulty gene on their X chromosome have. Females may develop night blindness or glare issues later in life, but rarely go blind from choroideremia.

But males have one X chromosome and one Y chromosome. They’re more prone to developing X-linked genetic disorders. Affected males pass on their X chromosome to their female children, who become carriers. Those female children have a 1 in 2 chance (50%) of passing the faulty X chromosome to their female children.

**SIGNS / SYMPTOMS**

The signs and symptoms of choroideremia include:

* Difficulty seeing at night (night blindness). This often happens before the age of 10.
* Difficulty with peripheral (side) vision.
* Lack of visual acuity (sharpness).
* Loss of the ability to see colors well.
* Increasing vision loss from the side that can progress to blindness.

**DIAGNOSIS METHOD**

It’s important to get regular eye exams, but an eye care provider will certainly examine your eyes if you report symptoms. They’ll likely do these tests, as well as others:

* Electroretinogram: This test measures your retina’s response to light. A provider will place electroencephalogram disks on your face, eyes, head and scalp, and then place very thin wires over the front of your eye, directly above your bottom eyelid. The technician will flash bright lights in front of your eyes to measure your retina’s activity.
* Optical coherence tomography (OCT): This type of test is a noninvasive imaging method that uses reflected light to create pictures of the back of your eye.
* Fluorescein angiography: Angiography is a test that uses dye injected through your veins to show the blood vessels in your eye.
* Genetic testing: A DNA test, or genetic testing, can identify mutations in your genes. A lab can use samples from your blood, hair, skin, tissue or amniotic fluid. Genetic testing can tell the difference between choroideremia and retinitis pigmentosa.

**TREATMENT OPTIONS**

Providers can’t cure choroideremia. But you may receive treatment for other issues. For instance, as vision gets worse, you may find that you can use some strategies for coping with low vision. Coping tips range from devices like magnifiers to counseling for mental health and genetics.

**PREVENTION TIPS**

Generally, people with choroideremia can expect their peripheral vision to get worse over time, leaving only central vision. Eventually, the condition may progress to blindness. There isn’t a cure.

**OUTLOOK / PROGNOSIS**

You can take care of yourself by following guidelines for a healthy life, including:

* Eating a healthy diet that includes green leafy vegetables, other vegetables, fruit and lean protein. Avoid excess salt, sugar and unhealthy fats.
* Getting enough physical activity.
* Quitting smoking if you smoke.
* Having regular eye exams.
* Getting enough sleep.

**WHEN TO SEE A DOCTOR**

You should see a healthcare provider whenever you have a change in vision. If any of the changes are sudden or painful, consider getting emergency care.

See a provider if you find you’re having difficulties with your daily activities. Ask for information on being eligible for gene therapy trials, too.

**DIFFERENTIAL DIAGNOSIS**

* Gyrate atrophy: Choroideremia in early stages may mimic gyrate atrophy of retina and choroid. Examination of fundus of family members, early presentation, and X-linked inheritance pattern are important features to clinically differentiate choroideremia from gyrate atrophy. Autosomal recessive, well demarcated scalloped areas of chorioretinal atrophy, nyctalopia in second to third decade, systemic hyperornithinemia, myopia and early cataracts
* Retinitis Pigmentosa: Waxy disc pallor, peripheral RPE bone spicule like degeneration, retinal arteriolar attenuation
* Myopic Degeneration: Absence of nyctalopia, tessellated fundus, lacquer cracks, diffuse atrophy, patchy atrophy, posterior staphyloma, high axial length, macular atrophy, straightened and stretched vessels, high rates of choroidal neovascularization, temporal peripapillary atrophy crescent, hemorrhages and tilting of the optic disc
* Ocular albinism: Infantile nystagmus, iris translucency, substantial hypopigmentation of ocular fundus, foveal hypoplasia, aberrant optic pathway projection associated with asymmetry of cortical responses on visual evoked potential testing
* Usher syndrome type 1: Autosomal recessive, pigmentary retinopathy, congenital deafness, imbalance from vestibular dysfunction
* Thioridazine hydrochloride retinal toxicity: History of medication use, loss of night vision, decreased ERG amplitudes, accumulation of fine or coarse pigment clumps, geographic RPE and choriocapillaris atrophyBietti crystalline dystrophy: Autosomal recessive, corneal deposits, yellow-white crystalline retinal deposits, progressive atrophy of the RPE, loss of choriocapillaris, progressive nyctalopia, visual field constriction, legal blindness in the fifth or sixth decades of life

**EPIDEMIOLOGY**

Choroideremia is a rare chorioretinal dystrophy that is estimated to affect between 1 in 50,000 to 1 in 100,000 individuals. Men are predominantly affected due to its X-linked etiology, but women can be asymptomatic carriers or rarely can be affected by the dystrophy as well. Northern Finland has the highest reported prevalence. There are thought to be more than 500 affected males in the United Kingdom and around 3000 throughout Europe.

*REFERENCES:* [**Choroideremia - EyeWiki**](https://eyewiki.org/Choroideremia)

[**Choroideremia: What It Is, Causes & Symptoms**](https://my.clevelandclinic.org/health/diseases/24569-choroideremia)

**COATS DISEASE**

**DEFINITION / DESCRIPTION**

Coats disease causes blood vessels in your child’s eye to develop incorrectly. Specifically, it affects blood vessels in their retina. Eventually, it makes the blood vessels swell and leak plasma into their retina. You might see it referred to as Coats’ syndrome or exudative retinitis.

The retina is the layer at the very back of your eyeball. It converts light that enters your eye into electrical signals your optic nerve sends to your brain, which creates the images you see.

Blood vessels are channels that carry blood throughout your body. They form a closed loop, like a circuit, that begins and ends at your heart.

Coats disease makes the tiny blood vessels in your child’s retina dilate (expand), twist and leak. The leak is usually small and takes time to cause symptoms you’ll notice — think about the difference between running over a nail and making your car’s tire lose air slowly and hitting a pothole and bursting it all at once.

Coats disease usually affects one eye (unilaterally), but it can develop in both eyes at the same time (bilateral Coats disease). It’s usually diagnosed in kids and teens but can develop at any age. It can make your child’s vision worse and lead to serious complications like a detached retina and blindness in their affected eye.

Talk to your healthcare provider or eye care specialist as soon as you notice any changes in your child’s eyes or vision.

**SIGNS / SYMPTOMS**

Your child might not have any symptoms of Coats disease, especially early on. If they do have symptoms, they might include:

* Worsening vision.
* Crossed eyes (strabismus).
* Eye pain.
* A white or silvery pupil (leukocoria).

**Coats disease stages**

Coats disease progresses (becomes more severe) in stages. They range from stage 1 (the mildest) to stage 5 (most severe):

* Stage 1 Coats disease: Your eye care specialist notices abnormal blood vessels in your child’s eye, but they’re not causing any issues or symptoms.
* Stage 2 Coats disease: The affected blood vessels in your child’s eye start leaking. This will probably affect your child’s vision.
* Stage 3 Coats disease: Your eye care specialist will classify the Coats disease as stage 3 if it causes a detached retina in your child’s affected eye.
* Stage 4 Coats disease: Stage 4 Coats disease causes glaucoma in your child’s eye.
* Stage 5 Coats disease: This is sometimes called end-stage Coats disease. It causes blindness in the affected eye.

**CAUSES**

Experts aren’t sure what causes Coats disease. It usually happens without a specific cause (idiopathically).

Coats disease isn’t a genetic disorder. This means it’s not passed through generations of a family — it’s not hereditary.

**RISK FACTORS**

Anyone can develop Coats disease. But boys are three times more likely to have it than girls.

Coats disease can develop at any age:

* Providers usually diagnose kids and teens before they’re 16.
* It’s rare, but babies can be born with Coats disease, and it can develop in some infants.
* Around one-third of people with Coats disease are adults older than 30.

**DIAGNOSIS METHODS**

An eye care specialist will diagnose Coats disease with an eye exam.

They’ll look at your child’s eyes (including inside them). They’ll examine your child’s eyes and vision with a visual acuity test.

Your eye care specialist might need one or a few imaging tests to diagnose Coats disease, including:

* Optical coherence tomography (OCT).
* Angiography.
* Ultrasound.
* MRI (magnetic resonance imaging).
* CT scan (computed tomography scan).

**Management and Treatment**

How Coats disease is treated depends on which stage your child has. If an eye care specialist diagnoses them early (stage 1 or early in stage 2), your child may not need any treatment. Your eye care specialist will regularly check your child’s eyes for changes in their retina’s blood vessels.

If your child has more severe symptoms or is in a later stage, Coats disease treatments include:

* Cryotherapy: Cryotherapy is a treatment where your healthcare provider applies extreme cold to freeze and destroy abnormal tissue. They’ll freeze the affected blood vessels in your child’s retina to prevent them from leaking anymore.
* Medication: An ophthalmologist can inject anti-vascular endothelial growth factor (VEGF) drugs into your child’s retina. This can stop their blood vessels from leaking and prevent a retinal detachment.
* Surgery: If your child has later-stage Coats disease or has experienced a detached retina, they’ll need surgery to repair the damage.

**PREVENTION TIPS**

Because it happens randomly with no cause, there’s nothing you can do to prevent Coats disease.

Visit your eye care specialist for follow-up eye exams to monitor any changes in your child’s retina after they’ve been diagnosed.

**OUTLOOK / PROGNOSIS**

How your child’s eye and vision will be affected by Coats disease depends on what stage it was diagnosed at, and how well their eye responds to treatment. Studies have found that the older kids are when they develop Coats disease, the less likely it is that they have severe cases that progress to later stages.

If Coats disease is diagnosed early in stages 1 or 2, there’s a good chance your child won’t have severe long-term effects. However, they might have reduced or low vision in their affected eye, even after treatment.

Later-stage Coats disease has a higher risk of blindness in the affected eye.

**Coats disease complications**

While your child has Coats disease, they’ll have an increased risk for other conditions that can affect their eye, including:

* Glaucoma.
* Cataracts.
* Uveitis.
* Neovascularization (developing new, abnormal blood vessels).

**WHEN TO SEE A DOCTOR**

See your healthcare provider or eye care specialist as soon as you notice any changes in your child’s eyes or vision.

Go to the emergency room if they have any of the following symptoms:

* A sudden loss of vision.
* Severe eye pain.
* They see new flashes or floaters in their eyes.

**Differential diagnosis**

1. Retinoblastoma
   * Most important and critical differential diagnosis, especially in children presenting with leukocoria.
   * Distinguishing features:
     + Retinoblastoma often presents bilaterally (though can be unilateral), while Coats disease is almost always unilateral.
     + Retinoblastoma shows intraocular calcifications detectable by ultrasound or CT, absent in Coats disease.
     + Retinal vessels in retinoblastoma course subretinally within the tumor mass, whereas in Coats disease telangiectatic vessels remain on the retinal surface.
     + Vitreous seeding is common in retinoblastoma but absent in Coats disease.
     + Retinoblastoma typically affects younger children (infants to toddlers), Coats disease often presents in older children or young adults.
2. Toxocariasis
   * Parasitic infection causing granulomatous inflammation and retinal lesions.
   * May present with leukocoria and retinal granulomas.
   * Serologic testing and history can aid differentiation.
3. Retinopathy of Prematurity (ROP)
   * Occurs in premature infants with abnormal retinal vascular development.
   * History of prematurity and characteristic peripheral retinal changes distinguish it.
4. Familial Exudative Vitreoretinopathy (FEVR)
   * Genetic disorder causing peripheral retinal avascularity and neovascularization.
   * Can present with exudation and retinal detachment similar to Coats disease.
   * Often bilateral and familial history may be present.
5. Pars Planitis (Intermediate Uveitis)
   * Inflammatory condition with vitreous cells and snowbanking.
   * Usually bilateral and associated with systemic symptoms.
6. Retinal Vasoproliferative Tumors
   * Peripheral retinal vascular masses with feeder and draining vessels, preretinal fibrosis, and extreme peripheral location.
   * Different from Coats disease’s telangiectatic vessels without a discrete mass.
7. Norrie Disease
   * X-linked recessive disorder causing bilateral retinal dysplasia and detachment.
   * Presents in infancy with blindness.
8. Eales Disease
   * Idiopathic retinal vasculitis with peripheral ischemia and neovascularization.
   * Usually affects young adults, often bilateral.
9. Cavernous Retinal Hemangioma
   * Vascular tumor with characteristic “cluster of grapes” appearance on fundus exam.
10. Leukemia and Other Retinal Metastases
    * May cause retinal infiltrates and hemorrhages mimicking exudation.
11. Persistent Fetal Vasculature (PFV)
    * Congenital anomaly causing persistent hyaloid vasculature and retinal detachment.

Diagnostic Tools to Differentiate Coats Disease

* Fundus Examination: Telangiectatic retinal vessels, aneurysms, and yellow subretinal exudates typical of Coats disease.
* Ultrasound and CT Scan: Absence of calcification favors Coats disease; calcifications suggest retinoblastoma.
* Fluorescein Angiography: Shows telangiectasia, aneurysms, leakage, and peripheral nonperfusion in Coats disease.
* Optical Coherence Tomography (OCT): Demonstrates retinal thickening, exudation, and detachment.
* MRI: Helpful in differentiating advanced Coats disease from retinoblastoma by showing subretinal exudates versus solid mass.

Other differential diagnoses of diseases with extensive hard exudates include

* Branch retinal venous occlusion,
* Vasoproliferative tumor,
* Capillary hemangioma,
* Chronic retinal detachment,
* Trauma, and
* Inflammation.

A Coats-like response (retinal telangiectasia, aneurysm, and hard exudates) may also be noted in

* Retinitis pigmentosa
* Healed choroiditis
* Tubercular subretinal abscess
* Retinal vasculitis,
* Pars planitis
* Leber Congenital amaurosis
* Facioscapulohumeral muscular dystrophy and deafness,
* Progressive facial hemiatrophy or Parry-Romberg syndrome or progressive hemifacial atrophy or hemifacial atrophy
* Isolated hemihyperplasia
* Tuberous sclerosis
* Alport's syndrome
* X-linked retinoschisis and regressed retinopathy of prematurity
* 'Blood-filled senile retinoschisis'
* Linear Scleroderma
* En coup de sabre scleroderma
* Epidermal nevus syndrome

**Epidemiology**

. The incidence was 0.09 per 100,000 population. All cases were unilateral, and 85% were male. The mean age at presentation was 146 months (median 96 months). Some authors have reported that the disease could be present at birth. In a large series, only 5% of cases were bilateral. The fellow eye was usually without symptoms and had subtle peripheral telangiectatic changes. Adult-onset Coats disease (first diagnosed at a minimum age of 35 years) constituted 7% of the 646 patients with Coats disease in a tertiary eye

REFERENCES

[Exudative Retinitis (Coats Disease) - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK560682/)

[Coats Disease: Symptoms, Stages & Treatment](https://my.clevelandclinic.org/health/diseases/coats-disease)

**Color Blindness**

**Definition and description**

Difficulty distinguishing colors due to retinal cone cell dysfunction. Color blindness is an eye condition in which someone can't see the difference between certain colors. Though many people commonly use the term "color blind" for this condition, true color blindness — in which everything is seen in shades of black and white — is rare. The medical term for color blindness is known as color vision deficiency.

Color blindness is usually inherited, meaning it's passed down through families. Men are more likely to be born with color blindness. Most people with color blindness can't tell the difference between certain shades of red and green. Less commonly, people with color blindness can't tell the difference between shades of blue and yellow.

Certain eye diseases and some medicines also can cause color blindness.

#### **Types of color blindness and what people can see**

There are several types of color blindness, defined according to which types of cones aren’t working well. To understand the types of color blindness, it helps to know a bit about cones.

Cones are nerve cells in your eye that detect colors in the visible spectrum of light. This spectrum includes all the wavelengths that humans can see. These range in length from 380 nanometers (short), or nm, to 700 nanometers (long). Normally, you’re born with three types of cones:

* Red-sensing cones (L cones): These cones perceive long wavelengths (around 560 nanometers).
* Green-sensing cones (M cones): These cones perceive middle wavelengths (around 530 nanometers).
* Blue-sensing cones (S cones): These cones perceive short wavelengths (around 420 nanometers).

Most people have all three types of cones, and these work as they should. However, if you have color vision deficiency, at least one type of cone isn’t working properly. Problems with your cones affect your ability to see colors in the traditional way. General categories that describe how many types of cones you have, and how well they’re working, include:

* **Trichromacy**: All three types of cones are present and working properly. You see all colors on the visible spectrum of light in the traditional way. This is full-color vision.
* **Anomalous trichromacy**: You have all three types of cones, but one type isn’t as sensitive to light in its wavelength as it should be. As a result, you don’t see colors in the traditional way, with variations from normal ranging from mild to severe. In mild cases, you may just confuse pale or muted colors. In more severe cases, you may also confuse vivid and pure (fully saturated) colors. These types of color blindness have names that end in “anomaly” (which indicates partial vision of a specific color).
* **Dichromacy**: One type of cone is missing. So, you only have two types of cones (usually S cones along with either L cones or M cones). You see the world through the wavelengths that those two types of cones can perceive. It’s hard to tell the difference between fully saturated colors. These types of color blindness have names that end in “anopia” (which indicates absence of vision of a specific color).
* **Monochromacy**: You only have one type of cone, or you have no cone function at all. You have very limited or no ability to see color. Instead, you see the world in varying shades of gray.

Within these general categories, there are many specific types of color blindness.

##### **Red-green color deficiency**

Red-green color deficiency is the most common type of color blindness. It affects how you see any colors or shades that have some red or green in them. There are four main subtypes:

* Protanopia: Your L cones are missing. So, you can’t perceive red light. You mostly see colors as shades of blue or gold. You may easily confuse different shades of red with black. You may also confuse dark brown with dark shades of other colors, including green, red or orange.
* Deuteranopia: Your M cones are missing. So, you can’t perceive green light. You mostly see blues and golds. You may confuse some shades of red with some shades of green. You may also confuse yellows with bright shades of green.
* Protanomaly: You have all three cone types, but your L cones are less sensitive to red light than they should be. Red may appear as dark gray, and every color that contains red may be less bright.
* Deuteranomaly: You have all three cone types, but your M cones are less sensitive to green light than they should be. You see mostly blues, yellows and generally muted colors.

Protanopia and deuteranopia are examples of dichromacy. Protanomaly and deuteranomaly are examples of anomalous trichromacy.

Other terms you might hear are “protan” and “deutan.” Protan and deutan are shorthand ways to talk about red-green colorblindness. Deutan refers to green (you have impaired or missing green-sensing cones, or M cones). Protan refers to red (you have impaired or missing red-sensing cones, or L cones).

Red-green color blindness is much more common among males. This is because the genes for the color vision cone light-sensitive proteins are on the X chromosome, of which males have one and females have two. So if the one X in a male contains abnormal genes, the color blindness will reveal itself, while females can compensate with the other normal gene on the second X chromosome.

##### **Blue-yellow color deficiency**

Blue-yellow color vision defects (tritan defects) are much less common and include:

* Tritanopia: You have no S cones. So, you can’t perceive blue light. You see mostly reds, light blues, pinks and lavender.
* Tritanomaly: You have all three cone types, but your S cones are less sensitive to blue light than they should be. Blues look green, and you see little or no yellow.

Blue-yellow color blindness equally affects males and females.

##### **Blue cone monochromacy**

This is the rarest form of color blindness. With this type, you don’t have working L cones or M cones. You only have S cones. It’s hard to tell the difference between colors, and you see mostly grays. You may also have other eye problems, including sensitivity to light (photophobia), nystagmus and nearsightedness.

##### **Rod monochromacy (achromatopsia)**

Achromatopsia is when all or most of your cones are missing or don’t work properly. You see everything in shades of gray. You also have other vision issues that may greatly impact your quality of life.

Inherited color blindness mostly affects males. This is due to its genetic inheritance pattern (X-linked recessive). Conditions passed down in this manner are much more common among males.

People can also acquire color blindness due to certain medical conditions, medications or environmental exposures

Among people of Northern European ancestry, red-green color blindness affects about 1 in 12 males and 1 in 200 females. These numbers vary by ethnicity. Some research shows that Europeans have the highest prevalence of color blindness.

Here are some statistics about the less common forms of color blindness:

* Blue-yellow color deficiency affects 1 in 10,000 people.
* Achromatopsia affects 1 in 30,000 people.
* Blue cone monochromacy affects 1 in 100,000 people.

Overall, around 300 million people around the world have some form of color blindness (mostly red-green).

**CAUSES**

Seeing colors across the light spectrum is a complex process that begins with your eyes' ability to respond to different wavelengths of light.

Light, which contains all color wavelengths, enters your eye through the cornea and passes through the lens and transparent, jellylike tissue in your eye (vitreous humor) to wavelength-sensitive cells (cones) at the back of your eye in the macular area of the retina. The cones are sensitive to short (blue), medium (green) or long (red) wavelengths of light. Chemicals in the cones trigger a reaction and send the wavelength information through your optic nerve to your brain.

If your eyes work as they should, you perceive color. But if your cones don't work properly, you will be unable to distinguish the colors red, green or blue.

**RISK FACTOR**

Several factors increase the risk of color blindness, including:

* **Gender.** Colorblindness is much more common in males than in females.
* **Family history.** Colorblindness is often inherited, meaning it is passed down through families. You can inherit a mild, moderate or severe degree of the condition. Inherited color deficiencies usually affect both eyes, and the severity doesn't change over your lifetime.
* **Diseases.** Some conditions that can increase the risk of color deficiency include sickle cell anemia, diabetes, macular degeneration, Alzheimer's disease, multiple sclerosis, glaucoma, Parkinson's disease, chronic alcoholism and leukemia. One eye may be more affected than the other, and the color deficiency may get better if the underlying disease can be treated.
* **Certain medicines.** Some medicines can affect color vision, such as hydrochloroquine, a medicine used to treat rheumatoid arthritis.
* **Damage to the eye.** Color blindness can be caused by trauma to the eye as a result of injury, surgery, radiation therapy or laser treatment.

**SIGNS AND** **SYMPTOMS**

You may have a color vision deficiency and not know it. Some people figure out that they or their child has the condition when it causes confusion — such as when there are problems differentiating the colors in a traffic light or interpreting color-coded learning materials.

People affected by color blindness may not be able to distinguish:

* Different shades of red and green.
* Different shades of blue and yellow.
* Any colors.

The most common color deficiency is an inability to see some shades of red and green. Often, a person who is red-green or blue-yellow deficient isn't completely insensitive to both colors. Defects can be mild, moderate or severe.

**DIAGNOSIS**

If you have trouble seeing certain colors, an eye care professional can test for a color deficiency. Testing likely involves a thorough eye exam and looking at specially designed pictures. These pictures are made of colored dots that have numbers or shapes in a different color hidden in them.

If someone has a color vision deficiency, they'll find it difficult or impossible to see some of the patterns in the dots.

**TREATMENT**

There are no treatments for most types of color vision difficulties, unless the color vision problem is related to the use of certain medicines or eye conditions. Stopping the medicine causing the vision problem or treating the underlying eye disease may result in better color vision.

Wearing a colored filter over eyeglasses or a colored contact lens may enhance perception of contrast between the confused colors. But such lenses won't improve the ability to see all colors.

### **Potential future treatments**

Some rare retinal disorders associated with color deficiency could possibly be modified with gene replacement techniques. These treatments are under study. One treatment was approved for a rare condition called Leber congenital amaurosis, a retina condition that is present at birth. More treatments might become available in the future.

**Lifestyle and home remedies**

Try the following tips to help you work around your color blindness.

* **Memorize the order of colored objects.** If it's important to know individual colors, such as with traffic lights, memorize the order of the colors.
* **Label colored items that you want to match with other items.** Have someone with good color vision help you sort and label your clothing. Arrange your clothes in your closet or drawers so that colors that can be worn together are near each other.
* **Use technology.** There are apps for phones and tablets that can help you identify colors.

## **Prevention**

You can’t prevent inherited color blindness. However, you may be able to lower your risk of acquired color blindness. Visit a healthcare provider for yearly checkups and ask about your risk for developing color vision deficiency. Some questions to ask include:

* Do any of my medical conditions put me at risk for color blindness?
* Can any of my medications cause color blindness?
* Should I be concerned about any chemical or environmental exposures at my job?
* What can I do to lower my risk?

## **Outlook / Prognosis**

Color blindness might not affect your life much, especially if your condition is mild. More severe forms may interfere with your job, education or personal life. It’s important to talk to your eye care provider about your condition and what you can expect going forward. If your child has color blindness, ask what you can do to help them with school.

### **Can color blindness affect my child’s career choice?**

Certain careers may be too challenging or unsafe to pursue with color deficiency. These include careers as an electrician, pilot, fashion designer or graphic artist. But you can encourage your child to pursue other careers where color vision won’t play a major role. Talk to counselors or mentors at your child’s school to access resources on career options.

## **Living With**

Connect with others who have color vision deficiency or parents of children with the condition. They can share advice and resources for living with color vision deficiency from day to day. Some tips include:

* Find a color buddy who can help with shopping for items like clothes or paint.
* Memorize the correct order of colors on things like traffic lights.
* Download apps that help you identify colors in the world around you.

**WHEN TO SEE THE DOCTOR**

If you suspect you have problems distinguishing certain colors or your color vision changes, see an eye doctor for testing. It's important that children get comprehensive eye exams, including color vision testing, before starting school.

There's no cure for inherited color deficiencies, but if illness or eye disease is the cause, treatment may improve color vision.

**Differential diagnosis (how it’s distinguished from other illnesses)**

## Congenital vs Acquired Causes

* Congenital Color Blindness:
  + Genetic defects affecting cone photopigments.
  + Most commonly red-green deficiencies inherited on the X chromosome.
* Acquired Color Vision Deficiency:
  + Due to ocular diseases (glaucoma, macular degeneration, diabetic retinopathy).
  + Neurological disorders (optic neuritis, multiple sclerosis).
  + Toxic or drug-induced (ethambutol, hydroxychloroquine).
  + Aging-related changes.

Other Conditions to Differentiate

* Visual Acuity Impairment: Poor vision can mimic color vision problems.
* Cerebral Achromatopsia: Cortical color blindness due to brain injury.
* Retinal Diseases: Affect cone function and color perception.
* Psychological or malingering causes: Rare but possible.

Diagnostic Tests

* Ishihara Color Plates: Screening for red-green deficiencies.
* Farnsworth-Munsell 100 Hue Test: Detailed color discrimination test.
* Anomaloscope: Gold standard for red-green color deficiency diagnosis.
* HRR (Hardy-Rand-Rittler) Test: Screens for red-green and blue-yellow deficiencies.
* Lantern Tests: Occupational screening for color discrimination.

### **Statistics**

* Color blindness is more common in white people.
* Red-green color deficiency is the most common form, occurring in about 1 in 12 males and 1 in 200 females with Northern European ancestry.
* Blue-yellow color vision deficiency is rarer and affects both males and females equally. This subtype presents in less than 1 in 10,000 people worldwide.
* Complete color blindness only affects around 1 in 30,000 people.
* One study found that 25% of people with anomalous trichromacy, in which one type of cone does not function correctly, were unaware that they had color vision deficiency.

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**COLOBOMA**

**Definition and description**

A coloboma is an area of missing tissue in your eye. Colobomas are present in a person’s eye when they’re born. They can affect one or both eyes.

The most recognizable and common colobomas affect your iris (the colored part of your eye) and cause your pupil (the dark center of your eye) to have a keyhole shape. But there are many types of colobomas, most of which you can’t see externally (on the outside).

Some colobomas cause no symptoms, but others can have serious impacts on your child’s vision. What symptoms they’ll have depends on where in their eye the coloboma developed and which kind of tissue they’re missing.

Your healthcare provider might be able to diagnose a coloboma when your baby is born.

Anyone can be born with a coloboma.

They’re a type of genetic disorder, which means they’re passed from parents to their children. You might see coloboma referred to as a congenital condition.

However, even if one parent (or both) has a coloboma, that doesn’t mean your child will definitely develop one.

Colobomas affect around 1 out of every 10,000 babies born each year.

Because not all colobomas cause noticeable symptoms, that number might be higher. Some people have an undiagnosed coloboma and never have symptoms or complications.

How much a coloboma affects your child depends on where in their eye it’s located. Colobomas can develop in one eye (unilateral coloboma) or both eyes (bilateral coloboma). Bilateral colobomas might affect different parts of your eyes.

Some colobomas won’t impact your child’s vision. Others cause total blindness in the affected eye. If your child has a coloboma on their retina, macula or optic nerve, they might have some vision, but it might be impaired.

You might not notice a coloboma affecting your child’s vision right away, especially while they’re very young.

People with colobomas might be more likely to have other issues with their eyes later in life, including:

· Cataracts.

· Glaucoma.

· Retinal detachment.

Colobomas can develop in almost any part of your child’s eye, including their:

· Iris.

· Retina.

· Optic nerve.

· Macula.

· Ciliary body.

· Eyelid.

### **Coloboma of the iris**

Colobomas of the iris are the most common type of coloboma. The iris is the colored part of your eye.

Babies born with a coloboma of the iris are sometimes referred to as having a congenital iris coloboma. Congenital is the medical term that means something is present at a person’s birth. Just like other types of colobomas, iris colobomas can be unilateral (only affecting one iris) or bilateral (affecting both irises). People with iris colobomas can also have other colobomas on their retina or optic nerve, too.

Colobomas of the iris are diagnosed and treated just like other types of colobomas. Some people with iris colobomas wear special contact lenses to help cover their pupils more completely. Surgery to change the appearance of an iris coloboma is an option, but not everyone is a good candidate for it. Talk to your provider or eye care specialist about which treatments will work for your child and when they should receive them.

A coloboma of the iris can give your child’s pupil a keyhole or cat-eye shape. The shape depends on where in their iris it is and how much tissue is missing.

It might look like their pupil is “leaking” into the colored part of their eye. Nothing is in danger of falling out or moving around in your child’s eye. Both the iris and pupil are solid layers of tissue. The missing piece of your child’s iris is simply revealing more of the pupil under it that would usually be covered.

A coloboma of the iris can affect your child’s vision.

Muscles in your iris control your pupil — the small black opening that lets light into your eye. Babies born with an iris coloboma are missing tissue in their iris. This can make it harder for their iris to control how wide (dilated) or narrow (contracted) their pupil is.

Depending on how big the coloboma is, it can cause symptoms that affect your child’s vision, including:

· Light sensitivity (photophobia).

· Blurry vision.

· Double vision (diplopia).

· “Ghost images,” blurry copies of an image you’re looking at or parts of an image that remain after you move your eyes.

**Causes**

Experts think a genetic disorder that affects a fetus’s eye while its developing during pregnancy causes colobomas.

Around two months before a baby is born, what’s known as the optic fissure comes together to form their eyes. When the fissure doesn’t completely close, it causes colobomas in one or both of your baby’s eyes.

Genes are made of DNA (deoxyribonucleic acid), which contain instructions for cell functioning and the characteristics that make you unique. Studies have shown links between certain genes in parents and the likelihood that their children will be born with a coloboma, but there isn’t enough evidence to say for sure which exact genes are responsible.

Certain external factors — like drinking alcohol during pregnancy — can increase the odds that your baby develops a coloboma.

**Signs and symptoms**

If your child has symptoms from their coloboma, they can include:

· Light sensitivity.

· A keyhole or cat-eye-shaped pupil.

· Low vision, blindness or partial vision loss.

· Nystagmus.

A coloboma might only affect a portion of your child’s field of vision (the full range of how much they can see). Colobomas can cause:

· Reduced peripheral vision.

· Difficulty with depth perception.

· A larger than usual blind spot.

**Diagnosis methods**

Your provider or an ophthalmologist will diagnose a coloboma during your child’s eye exam. They’ll examine your child’s eye and look inside it to identify any missing tissue. Every person with a visible coloboma (one you can see on the outside) should have a dilated eye exam to check for other colobomas inside their eye.

## **Management and Treatment**

There isn’t a treatment to replace the missing tissue in your child’s eyes.

However, there are treatments that might improve their vision, including:

· Wearing corrective lenses (glasses or contacts).

· Wearing an eye patch to prevent a lazy eye (amblyopia).

· Low vision aids (if their vision can’t be improved with corrective lenses).

Some people with iris colobomas can have surgery to change the appearance of their affected eye.

## **Prevention**

You can’t prevent genetic conditions like colobomas from developing during your pregnancy.

Colobomas — and other genetic conditions — are linked to certain environmental factors (things that happen to or around a person who is pregnant), including:

· Drinking alcohol.

· Smoking or using tobacco products.

· Using recreational drugs.

Talk to your provider about what you should avoid eating, drinking or doing while you’re pregnant.

## **Outlook / Prognosis**

How much a coloboma affects your child’s life depends on where it is in their eye. Many people with colobomas never have any symptoms and can live their whole lives without any complications. Other people’s vision is affected from birth.

Even if a coloboma impairs your child’s sight, it isn’t fatal and can’t spread.

Very rarely, a coloboma is a sign of what’s known as CHARGE syndrome — a genetic syndrome that can be life-threatening. Talk to your provider or ophthalmologist about your baby’s risk and what you and your child should expect as they grow and develop.

## **Living With**

### **When should I see my healthcare provider?**

See your healthcare provider as soon as you notice any changes in your child’s eyes or vision.

Go to the emergency room if your child has any of the following symptoms:

· A sudden loss of vision.

· Severe eye pain.

· Sees new flashes or floaters in their eyes.

### **What questions should I ask my doctor?**

· What type of coloboma does my child have?

· How will this affect their vision?

· What treatments are available?

· Do they have a higher risk for other issues in their eyes?

**DIFFERENTIAL DIAGNOSIS**

If there is an absence of eyelid tissue, trauma to the tissue should be considered on the differential and ruled out. If the defect has been present at birth, it is most likely a coloboma. The clinician must consider the following syndromes and investigate if they establish the diagnosis of an eyelid coloboma.

1. Fraser syndrome
2. Goldenhar syndrome
3. Treacher Collins syndrome (mandibulofacial dysostosis)
4. CHARGE syndrome
5. Frontonasal dysplasia
6. Delleman-Oorthuys (oculocerebrocutaneous) syndrome
7. Nasopalpebral lipoma coloboma syndrome
8. Manitoba oculotrichoanal (MOTA) syndrome
9. Anophthalmia and micophthalmia

**EPIDEMIOLOGY**

Eyelid coloboma is estimated to occur in approximately 1 in 10,000 births, with no gender predilection. Eyelid coloboma does not exhibit racial predisposition, except for the Manitoba oculotrichoanal syndrome, which is specific to the aboriginal population of northern Manitoba. One study found bilateral eyelid coloboma in 44.4% of cases. Goldenhar syndrome is strongly associated with eyelid coloboma and occurs in about 1 in 5600 births.

Eyelid coloboma is estimated to occur in approximately 1 in 10,000 births, with no gender predilection.

Eyelid coloboma does not exhibit racial predisposition, except for the Manitoba oculotrichoanal syndrome, which is specific to the aboriginal population of northern Manitoba. Bilateral eyelid coloboma is found in 44.4% of cases.

Notably, Goldenhar syndrome is strongly linked to eyelid coloboma and occurs in about 1 in 5,600 births.

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**Conjunctivitis (Also known as Pink Eye)**

**Definition and description**

Conjunctivitis (Pink Eye) is an infectious disease where the whites of the eye turn pink due to an inflammation of the conjunctiva which is mostly due to an infection, allergy, or irritants, causing redness and may have a sticky discharge. It can be treated with medications.

**Causes and risk factors**

Causes of pink eye include:

* Viruses.
* Bacteria.
* Allergies.
* A chemical splash in the eye.
* A foreign object in the eye.
* In newborns, a blocked tear duct.

### **Viral and bacterial conjunctivitis**

Most cases of pink eye are caused by adenovirus. It also can be caused by other viruses, including herpes simplex virus and varicella-zoster virus.

Both viral and bacterial conjunctivitis can occur along with colds or symptoms of a respiratory infection, such as a sore throat. Wearing contact lenses that aren't cleaned properly or aren't your own can cause bacterial conjunctivitis.

Both types are very contagious. They are spread through direct or indirect contact with the liquid that drains from the eye of someone who's infected. One or both eyes may be affected.

### **Allergic conjunctivitis**

Allergic conjunctivitis affects both eyes. It is a response to an allergy-causing substance such as pollen. In response to allergens, your body produces an antibody called immunoglobulin E (IgE). IgE triggers special cells in the mucous lining of your eyes and airways to release inflammatory substances, including histamines. Your body's release of histamine can produce a number of allergy symptoms, including red or pink eyes.

If you have allergic conjunctivitis, you may experience intense itching, tearing and inflammation of the eyes. You also could have sneezing and watery nasal discharge. Most allergic conjunctivitis can be controlled with allergy eye drops. Allergic conjunctivitis is not contagious.

### **Conjunctivitis resulting from irritation**

Irritation from a chemical splash or foreign object in the eye also is associated with conjunctivitis. Sometimes flushing and cleaning the eye to wash out the chemical or object causes redness and irritation. Symptoms, which may include watery eyes and a mucous discharge, usually clear up on their own within about a day.

If flushing doesn't resolve the symptoms, or if the chemical is a caustic one such as lye, see your healthcare professional or eye specialist as soon as possible. A chemical splash into the eye can cause permanent eye damage. Ongoing symptoms could indicate that you still have the foreign body in your eye. Or you also could have a scratch on the cornea or the membrane covering the eyeball, called the conjunctiva.

**RISK FACTOR**

Risk factors for pink eye include:

* Exposure to someone infected with the viral or bacterial form of conjunctivitis.
* Exposure to something you're allergic to, for allergic conjunctivitis.
* Using contact lenses, especially extended-wear lenses.

**Signs and symptoms**

The most common pink eye symptoms include:

* Redness in one or both eyes.
* Itchiness in one or both eyes.
* A gritty feeling in one or both eyes.
* A discharge in one or both eyes that forms a crust during the night that may prevent your eye or eyes from opening in the morning.
* Tearing.
* Sensitivity to light, called photophobia.

**Diagnosis methods**

In most cases, your healthcare professional can diagnose pink eye by asking about your recent health history and symptoms and examining your eyes.

Rarely, your care professional may take a sample of the liquid that drains from your eye for laboratory analysis, called a culture. A culture may be needed if your symptoms are severe or if your care professional suspects a high-risk cause, such as:

* A foreign body in your eye.
* A serious bacterial infection.
* A sexually transmitted infection.

**Treatment options**

Pink eye treatment is usually focused on symptom relief. Your care professional may recommend:

* Using artificial tears.
* Cleaning your eyelids with a wet cloth.
* Applying cold or warm compresses several times daily.

If you wear contact lenses, you'll be advised to stop wearing them until treatment is complete. Your care professional will likely recommend that you throw out soft contacts you've already worn.

Disinfect hard lenses overnight before you reuse them. Ask your care professional if you should discard and replace your contact lens accessories, such as the lens case used before or during the illness. Also replace any eye makeup used before your illness.

In most cases, you won't need antibiotic eye drops. Since conjunctivitis is usually viral, antibiotics won't help. They may even cause harm by reducing their effectiveness in the future or causing a medicine reaction. Instead, the virus needs time to run its course. This typically takes around 2 to 3 weeks.

Viral conjunctivitis often begins in one eye and then infects the other eye within a few days. Your symptoms should gradually clear on their own.

Antiviral medicines may be an option if your viral conjunctivitis is caused by the herpes simplex virus.

### **Treatment for allergic conjunctivitis**

If the irritation is allergic conjunctivitis, your healthcare professional may prescribe one of many different types of eye drops for people with allergies. These may include medicines that help control allergic reactions, such as antihistamines and mast cell stabilizers. Or your care professional may recommend medicines to help control inflammation, such as decongestants, steroids and anti-inflammatory drops.

Nonprescription versions of these medicines also may be effective. Ask your care professional about the best option for you.

You might lower the severity of your allergic conjunctivitis symptoms by avoiding whatever causes your allergies.

**Lifestyle and home remedies**

To help you cope with the symptoms of pink eye until it goes away, try to:

* **Apply a compress to your eyes.** To make a compress, soak a clean, lint-free cloth in water and wring it out before applying it gently to your closed eyelids. Generally, a cool water compress will feel the most soothing. You also can use a warm compress if that feels better to you. If pink eye affects only one eye, don't touch both eyes with the same cloth. This reduces the risk of spreading pink eye from one eye to the other.
* **Try eye drops.** Nonprescription eye drops called artificial tears may relieve symptoms. Some eye drops contain antihistamines or other medicines that can help people with allergic conjunctivitis.
* **Stop wearing contact lenses.** If you wear contact lenses, you may need to stop wearing them until your eyes feel better. How long you'll need to go without contact lenses depends on what's causing your conjunctivitis. Ask your healthcare professional whether you should throw away your disposable contacts, as well as your cleaning solution and lens case. If your lenses aren't disposable, clean them thoroughly before reusing them.

**Prevention tips**

Practice good hygiene to control the spread of pink eye. For instance:

* Don't touch your eyes with your hands.
* Wash your hands often.
* Use a clean towel and washcloth daily.
* Don't share towels or washcloths.
* Change your pillowcases often.
* Throw away old eye cosmetics, such as mascara.
* Don't share eye cosmetics or personal eye care items.

Keep in mind that pink eye is no more contagious than the common cold. It's okay to return to work, school or child care if you're able to practice good hygiene and avoid close contact. However, if work, school or child care involves close contact with others, it may be best to stay home until your or your child's symptoms clear up.

### **Preventing pink eye in newborns**

Newborns' eyes are susceptible to bacteria present in the mother's birth canal. These bacteria often cause no symptoms in the mother. In some cases, these bacteria can cause infants to develop a serious form of conjunctivitis known as ophthalmia neonatorum. This condition needs immediate treatment to keep the baby's sight. That's why shortly after birth, an antibiotic ointment is applied to every newborn's eyes. The ointment helps prevent eye infection.

**Prognosis**

The outlook for pink eye is generally good, especially when treated as needed. Milder cases often go away on their own with no treatment.

If you notice treatments aren’t working to help your pink eye, call the provider treating you. They may be able to adjust your treatment to better help you.

#### **How long does pink eye last?**

Pink eye can have different expected timelines, depending on the type you have. Allergy-related pink eye lasts as long you’re around the allergen causing the reaction. Bacterial infections last up to 10 days (and fewer when treated). Viral infections typically last up to two weeks, but some may last longer in rare cases.

If you suspect you have pink eye and it keeps worsening after a few days, it’s a good idea to see a primary care provider or go to urgent care.

You or your child can usually go back to daycare, school or work as soon as symptoms go away. This might be as soon as 24 hours after antibiotic treatment for a bacterial infection and between two and seven days after a viral infection.

Being symptom-free means you don’t have:

* Yellowish discharge.
* Crusting on your eyelashes or in the corners of your eyes.
* Pink color.

Be sure to check with your healthcare provider about when it’s safe to return. If an allergy or something else that’s not contagious caused your pink eye, you don’t need to stay home.

**Possible complications**

In both children and adults, pink eye can cause inflammation in the cornea that can affect vision. Prompt evaluation and treatment by your healthcare professional can reduce the risk of complications. See your professional if you have:

* Eye pain.
* A feeling that something is stuck in the eye.
* Blurred vision.
* Light sensitivity.

**When to see a doctor / red flag**

There are serious eye conditions that can cause eye redness. These conditions may cause eye pain, a feeling that something is stuck in your eye, blurred vision and light sensitivity. If you experience these symptoms, seek urgent care.

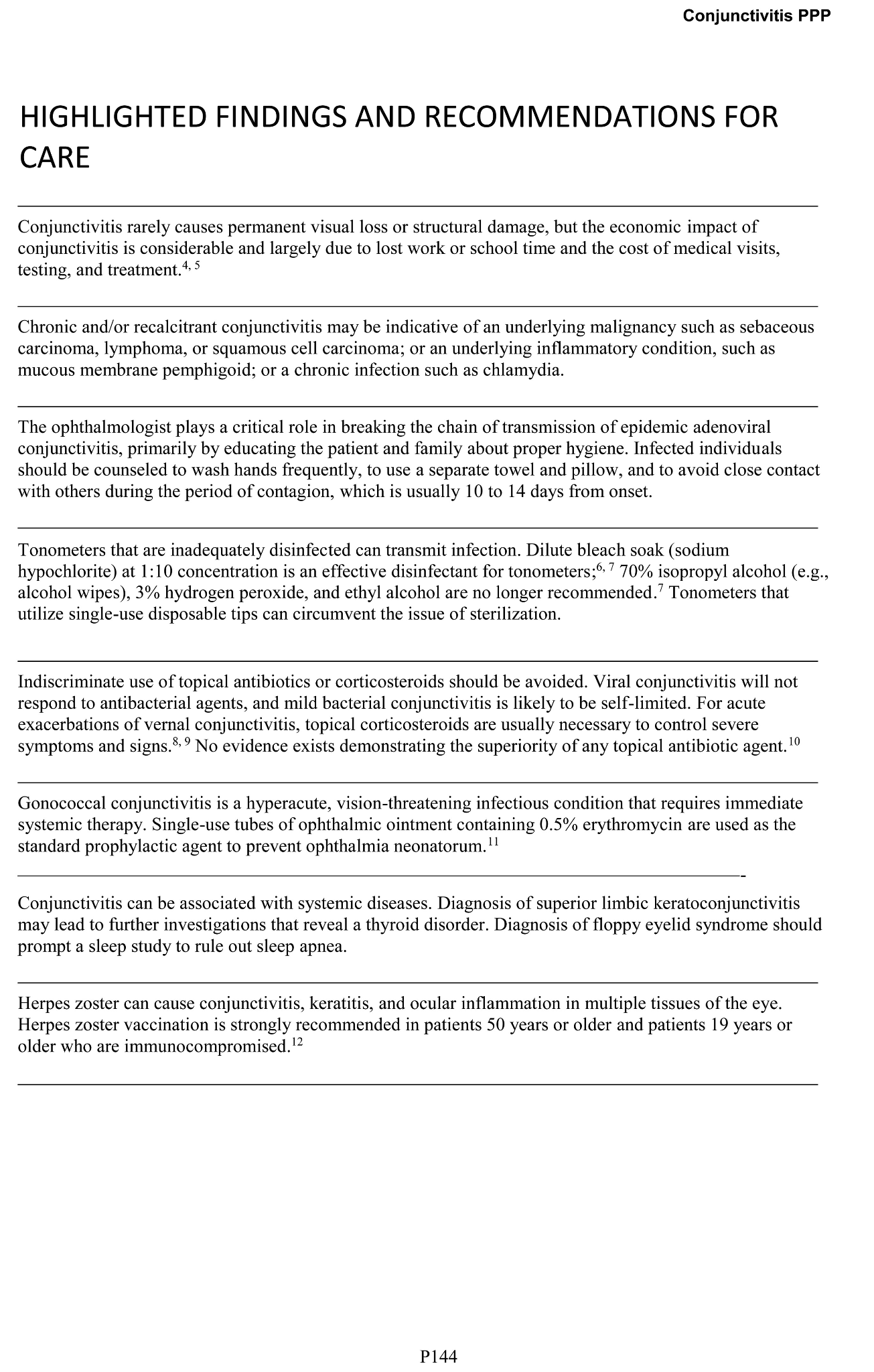
People who wear contact lenses need to stop wearing their contacts as soon as pink eye symptoms begin. If your symptoms don't start to get better within 12 to 24 hours, make an appointment with your eye healthcare professional to make sure you don't have a more serious eye infection related to contact lens use.

**Differential diagnosis**

There are many emergent and non-emergent causes of eye redness. When diagnosing conjunctivitis, ruling out the emergent causes that can lead to vision loss is essential. The differentials for conjunctivitis include:

* Glaucoma
* Iritis
* Keratitis
* Episcleritis
* Scleritis
* Pterygium
* Corneal ulcer
* Corneal abrasion
* Corneal foreign body
* Subconjunctival hemorrhage
* Blepharitis
* Hordeolum
* Chalazion
* Contact lens overwear
* Dry eye

**Recent guidelines or updates**



**Epidemiology data**

The occurrence of conjunctivitis depends on various factors such as age, gender, and time of the year. In the emergency department, cases of acute conjunctivitis show a bimodal distribution. The first peak is observed among children under 7, with the highest incidence between 0 and 4 years. The second peak occurs at 22 years in women and 28 years in men. Though overall rates of conjunctivitis diagnosed in the emergency department are slightly higher in women than in men, seasonality also plays a role in the presentation and diagnosis of conjunctivitis. Across all age groups, there is a peak incidence of conjunctivitis in children 0 to 4 years in March, followed by other age groups in May.

Regardless of changes in climate or weather patterns, seasonality is consistent for all geographic regions, as described in a nationwide emergency department study. Allergic conjunctivitis is the most common cause of conjunctivitis, affecting 15% to 40% of the population, and is often observed in spring and summer. Bacterial conjunctivitis rates are highest from December to April. Allergic conjunctivitis is considered the most common allergic ocular disease, affecting 15% to 20% of the population, with seasonal and perennial types

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**Corneal disease and dystrophies/ ULCER**

**Definition and description**

### There are several common causes of corneal disease, including infections, trauma, allergies, and more.The cornea is the clear tissue at the front and center of the eye. Its transparency permits light to pass into the eye, through the pupil, lens, and onto the retina at the back of the eye. The three major corneal layers are the outer layer of the cornea or epithelial layer, the middle layer termed the stroma, and finally, a single layer of cells called the endothelium.

### The curvature of the cornea plays an important role in focusing (refracting or bending) light. The normal cornea is smooth, clear, and tough. It helps protect the eye from infection and foreign material.

### There are several common causes of corneal disease, including the following:

### Infections

### Bacterial, fungal, or viral keratitis, as well as parasitic diseases

### Trauma

### Abrasions or exposure to toxic chemicals

### Dystrophies and degenerative corneal disorders

### Fuchs' dystrophy, map-dot-fingerprint dystrophy, or lattice corneal dystrophy

### Autoimmune disorders

### Wegener's disease, rheumatoid arthritis, or lupus

### Nutritional deficiencies

### Vitamin A deficiency

### Allergies

### Vernal and atopic keratoconjunctivitis

### Growths

### Pterygium or benign or malignant cancerous growths on the eye's surface

### Ectasia (thinning)

### Keratoconus, or thinning of the cornea following refractive laser surgery

### Stevens-Johnson syndrome is a rare but serious inflammatory reaction to a medication or an infection

### The cornea can also be damaged secondarily by other common eye conditions such as tear film abnormalities (dry eye), eyelid disorders, glaucoma, and iridocorneal endothelial syndrome (ICE), which may be associated with glaucoma.

### 

### **Causes and risk factors of corneal disease**

The causes of corneal disease vary widely. The conditions listed above are due to hereditary (inherited) causes, infection, trauma, autoimmune disorders, nutritional deficiencies, allergy, secondary causes (other eye diseases that also affect the cornea), growths, and tumors among others.

### **What causes corneal ulcers?**

Corneal ulcers can happen for many reasons, but they mainly break down into two categories: infectious and noninfectious.

#### **Infectious causes**

Infectious causes are conditions that you can catch from or spread to others. There are four subtypes of infectious causes:

* Bacteria. The most common types of bacteria that infect the cornea are *Pseudomonas*, *Staphylococcus* and *Streptococcus* species. Many of these bacteria are present all around us but cause infections if the cornea has an injury.
* Viruses. These include common viruses like Herpes simplex viruses (which cause cold sores or genital herpes) and varicella-zoster virus (which causes both chickenpox and shingles).
* Fungi. These are most likely with eye injuries from plants or soil (like while gardening. The most common fungi that do this include *Aspergillus* and *Candida*. Fungal infections are also a more serious concern for people who are immunocompromised (their immune systems are suppressed). Their immune systems can’t fight off infections well, so those fungi can establish themselves more easily.
* Parasites. *Acanthamoeba* family parasites are the most common cause of parasitic infection-related corneal ulcers. These parasites are a major risk for people who swim while wearing contacts. That applies to both bodies of water or pools because *Acanthamoeba* species can survive lower levels of chlorination (like in undertreated swimming pools). They can also live in tap water, which is why you should never use tap water to clean or store your contacts.

#### **Noninfectious causes**

These are conditions or circumstances that can cause corneal ulcers without an infection. Examples include:

* Eye injuries. Burns, scratches (corneal abrasions), cuts (lacerations) and punctures can all lead to ulcers when they don’t heal correctly. They also make your eyes more vulnerable to infections, which can lead to corneal ulcers forming.
* Exposure. If you can’t close your eyes fully (a condition called lagophthalmos), that leaves your corneas exposed for much longer than they should be. This can lead to corneal surface damage. Your corneas are also vulnerable to exposure damage in very hot or cold conditions.
* Very dry eyes. This can be because of weather conditions, eye conditions or a combination of the two.
* Toxic effects. These can be from toxic substances or, more rarely, from medications you’re taking.
* Immune conditions. Sometimes, eye inflammation happens because your immune system malfunctions. That inflammation can weaken your corneal tissue, making it vulnerable to damage and ulcer formation.

Risk factors similarly vary depending on the individual's circumstances. Some risk factors are not modifiable, such as inherited genetic conditions. Others might be avoidable by limiting exposure to trauma and infection. In many people, prompt treatment of corneal disease in its early stages will minimize the severity of the disease and its complications.

### **Risk factors for this condition**

Anyone can get corneal ulcers, but you have a higher risk if you have:

* Contact lenses that you wear often or for long periods (especially if you sleep or swim with them still in, or don’t maintain them properly).
* A current or past herpes simplex virus infection or varicella-zoster virus infection.
* Dry eyes.
* Conditions that make it harder or impossible to fully close your eyes (including different types of facial paralysis like Bell’s palsy, or conditions that make your eyes bulge, like Graves’ disease).
* Steroid-containing medicated eye drops that you’re currently using or recently used.
* An injury or burn on your cornea.
* Type 2 diabetes.
* A history of eye surgery.
* A history of other eye diseases, especially corneal diseases.

### **Signs and symptoms of corneal disease**

Signs of corneal problems can include redness around the cornea and/or corneal cloudiness.

Symptoms include the following:

* visual impairment, such as blurred or cloudy vision,
* severe pain in the eye,
* tearing, and
* sensitivity to light.
* Some patients have additional symptoms of headache, nausea, and fatigue.

Symptoms of a corneal ulcer include:

* pain,
* intense redness,
* feeling as if the eye is scratched or something is in the eye,
* sensitivity to light, and
* blurry vision.

Blurred vision may be the result of an irregular tear layer or epithelial layer (as seen in the dry eye), scarring (following trauma or infection), cataracts, deformity of the corneal curvature (as seen in keratoconus), or swelling of the cornea (as seen in Fuchs' dystrophy). Pain and light sensitivity can be quite severe, especially in conditions affecting the outermost layer (epithelium) of the cornea. Examples include traumatic abrasions, infectious ulcers, and erosions from dryness.

If you suspect a corneal ulcer or have the symptoms of a corneal ulcer and wear contact lenses, see your ophthalmologist immediately. High potency antibiotics and pain medications are the treatments for this condition.

Corneal ulcer symptoms vary widely. The most common ones include:

* Red or bloodshot eyes.
* Teary or watery eyes (epiphora).
* Eye pain (can vary from mild discomfort or aching to severe pain).
* Feeling like something’s stuck in your eye, like a hair or dust (foreign object sensation).
* Light sensitivity (photophobia).
* Blurred vision.
* Inflamed or swollen eyelids (blepharitis).
* A white or gray spot on your cornea (these aren’t always visible, and even when they are, they can be hard to see without the right tools).

**Diagnosis methods**

To diagnose a corneal ulcer, an eye specialist or another healthcare provider will rely mainly on an eye exam. Unlike a routine exam that checks all parts of your eye health, these exams will be more issue-specific. A key part of the eye exam that they’ll use is the slit lamp exam. It lets your eye specialist get an up-close, detailed look at your corneas.

One particular lab test your provider might do is a swab culture. That involves taking a soft-tipped swab, collecting some of the discharge from your eye and then sending that to a lab for testing. The test results may tell your provider what kind of infection is causing your ulcer. In cases that aren’t responsive to treatment, your provider may recommend taking this a step further and doing a corneal biopsy. That involves taking a sample of your corneal tissue for testing.

Your eye specialist can tell you more about any other tests they recommend. They can also explain how the tests work, why they should help and any side effects you should expect after the tests.

An eye doctor will review the person's medical history and perform a careful examination of the eyes and eyelids. The cornea is examined in detail using a slit lamp microscope.

Additional medical testing that can provide the information needed to make a diagnosis may include:

* Topography and keratometry (to study the shape of the cornea)
* Pachymetry (to measure the thickness of the cornea)
* Specialized microscopy (providing detailed pictures to assess the health of the endothelial cells, or to identify infectious agents)
* Assessment of the tear film

In some individuals, cultures, biopsies, or blood tests are also necessary.

## **Management and Treatment**

Corneal ulcers can happen for many reasons, which means there are many ways to treat them. Some treatments are very specific and only work for certain conditions. Other treatments are more general, helping the symptoms that are most likely with corneal ulcers.

Some of the more common treatment options include:

* Medications. These can include antibiotics for bacterial infection-related ulcers or other similar approaches for viruses, fungi and parasites. Other medications, like corticosteroids, are more general and treat symptoms of corneal ulcers. Your provider may also recommend nonsteroidal anti-inflammatory drugs (NSAIDs) like ibuprofen (Advil® or Motrin®). Your eye specialist can tell you about the medications, the form they come in and more.
* Scleral lenses or bandage contact lenses. These are wearable items that help protect your eyes from further damage, giving your corneas time to heal.
* Tarsorrhaphy. This treatment involves keeping your eye shut for longer periods. To do that, your provider can physically suture your eye closed, or they can use medications like onabotulinumtoxinA (Botox®) to temporarily paralyze the muscles that control your eyelids. Keeping your eye shut gives your cornea a more ideal environment for healing.
* Surgery. In cases where corneal ulcers aren’t healing, you may need surgery. In more severe cases, a corneal transplant might be necessary.

Other treatments may be possible or necessary, depending on your specific circumstances. Your eye specialist is the best source of information about the treatment options available for your specific case. They can also tell you more about the possible side effects or complications from treatment.

### **What is the treatment for corneal disease?**

Treatment is tailored to the individual disease and the individual patient. Treatments might include medications, laser treatment, or surgery, depending on the condition.

Infections are treated with medicated eye drops (antibiotics, antivirals, and antiparasitics) and, in some cases, oral medication. Herpetic stromal keratitis is a recurring swelling that develops after a herpes eye infection and is managed with anti-inflammatory steroid eye drops.

An abrasion might require temporary patching or a bandage contact lens, depending on the cause and extent of the injury.

Keratoconus, in which the cornea can take on a distorted cone shape, is often managed with special contact lenses. Newer treatments, including corneal crosslinking (riboflavin and ultraviolet-A) and corneal implants, are also options. Advanced keratoconus diseases are treated with anterior lamellar keratoplasty for corneal transplant surgery.

Chronic swelling from Fuchs' dystrophy or other conditions that damage the cornea's endothelial cells is managed initially with salty eye drops or ointments that help prevent the accumulation of fluid within the cornea. If the condition worsens, an endothelial lamellar keratoplasty (a type of partial-thickness transplant surgery) may be indicated.

Research is underway to develop an artificial cornea for transplantation.

Autoimmune disorders are best treated by addressing the underlying disease. Corneal involvement is often managed with anti-inflammatory eye drops such as steroids; however, steroid-sparing immune-modulating medications are sometimes preferable, particularly when other parts of the body are also involved.

Eye problems caused by vitamin A deficiency, which can be seen in patients who have had certain types of bariatric (weight loss) surgery, can be corrected with supplements.

Allergic eye disease responds well to both topical and oral allergy medication.

A pterygium is a growth on the cornea's surface; this is most commonly seen after chronic sun exposure. They can be removed surgically if they become bothersome. Cancers of the surface of the eye are managed with surgery or in some cases, topical chemotherapy eye drops or injections.

A dry eye is common and can result in painful erosions of the corneal surface. Aside from lubricating the eyes with artificial tears, addressing the underlying cause is important. In some individuals, dryness is due to a lack of tear production, and anti-inflammatory drops such as cyclosporine (Restasis) or steroids may help. In other cases, the dryness is due to the evaporation of the tears between blinks. This occurs when the eyelids' oil glands (Meibomian glands) are not functioning well. Normally, the oil from these glands coats the eye's surface and prevents tear evaporation. The oil glands' function can be improved with a combination of warm compresses, lid hygiene (for example, dilute baby shampoo lid scrubs), increased intake of omega-3 fatty acids, and in some patients, oral medication.

**Prevention tips of corneal disease**

Many corneal diseases are preventable by reducing risk factors. For example, maintaining optimal eye health (with good hygiene and regular vaccinations) is the best prevention against many infectious diseases. There are vaccines available to reduce the severity and frequency of shingles, which can result in an eye herpes infection called herpes zoster ophthalmicus.

Contact lens wear can make individuals especially susceptible to serious corneal infections, so people should clean contact lenses as directed. Glasses and sunglasses with 100% ultraviolet block can protect against growths that are associated with sun exposure, such as pterygium, and eye surface cancers. Safety glasses should be worn when warranted to prevent trauma. A diet rich in omega-3 fatty acids and sufficient vitamin A may help maintain a healthy tear film, thus minimizing dry eye symptoms.

Reviewing one's family's ocular health history helps look for hereditary conditions. Regular eye examinations are important for detecting eye diseases at their earliest stages.

A corneal ulcer is a common eye condition. It refers to a small crater (ulcer) on the front part of the eye, usually following an infection. Bacteria, viruses, or fungus can cause a corneal ulcer.

People who wear contact lenses are at higher risk for corneal ulcers. That’s because infectious agents may get trapped behind a lens. People deficient in vitamin A are particularly vulnerable to corneal ulcers.

Corneal ulcers aren’t 100% preventable, but there are many things you can do to lower your odds of developing them. Some things you can do include:

* Don’t sleep in contact lenses. Sleeping in contact lenses is the most common cause of serious corneal ulcers.
* Don’t wear contact lenses beyond their lifespan. The longer you use soft contact lenses, the more likely they are to grow bacteria. Daily contact lenses are the safest.
* Wash or sanitize your hands frequently. Use soap and water if your hands look or feel dirty. If they don’t look or feel dirty, you can use an alcohol-based hand sanitizer (at least 60% alcohol).
* Use eye protection. Protecting your eyes from injury is a key way to avoid injury-related ulcers. Make sure you use the right protection for your eyes, too.
* Never share things that touch your eyes. This goes for any items that touch areas of your face around your eyes. Some common examples include makeup or hygiene items like washcloths or towels. It also includes items for storing things that touch your eyes, like contact lens cases.

## **Outlook / Prognosis**

Corneal ulcers can vary widely depending on what causes them. That includes how long they take to heal or how well they should heal. Your eye specialist can tell you more about what you can expect with your specific case.

Some key factors that can impact your outlook are:

* Size. Larger ulcers usually have a less favorable outlook.
* Cause. Some causes are very difficult to treat or recover from.
* Location. Where an ulcer happens on your cornea can affect the outcome.
* Response to treatment. The outlook is usually better when ulcers respond well to treatment.

#### **How long do corneal ulcers take to heal?**

The healing time for corneal ulcers can vary widely, and many factors play a role in this. Your eye specialist is the best person to tell you the likely time frame for your case, as they can tailor the estimate to the details of your situation.

## **Living With**

If you have a corneal ulcer, the most important things you can do include:

* Follow your provider’s instructions on treatment closely. That goes for medications, how you use or protect your eye, etc.
* See your provider as recommended. Follow-up visits let your provider monitor an ulcer’s healing and adjust treatment if necessary.
* Call your eye specialist if you have questions. That includes if you have any new symptoms, questions about how to take medications or treatments, or if you notice treatments aren’t effective. You should also talk to your specialist if any side effects from treatment are causing issues. They may have ways to reduce the impact of symptoms or adjust your treatment to stop side effects from happening.

### **What are the potential complications of corneal disease?**

Many corneal diseases are treatable and have a good prognosis. However, vision loss and chronic eye pain are potential complications of corneal disease so it is important to review treatment options carefully with an eye doctor.

### **What are the complications of corneal ulcers?**

Corneal ulcers can cause the following complications:

* Astigmatism or other vision changes.
* Cataracts.
* Endophthalmitis.
* Glaucoma.
* Perforated or scarred cornea.
* Recurrent corneal erosions (RCEs).
* Vision loss.

**Differential diagnosis**

Infectious Corneal Diseases

* Bacterial Keratitis: Usually presents with pain, redness, discharge, and corneal ulceration. Common pathogens include *Staphylococcus aureus*, *Pseudomonas aeruginosa*.
* Viral Keratitis: Often caused by herpes simplex virus (HSV), characterized by dendritic ulcers and recurrent episodes. Varicella-zoster virus can cause herpes zoster ophthalmicus with keratitis.
* Fungal Keratitis: Occurs after trauma with vegetative matter; presents with feathery-edged infiltrates.
* Acanthamoeba Keratitis: Associated with contact lens wear; severe pain disproportionate to clinical signs.

2. Non-Infectious Inflammatory Corneal Diseases

* Keratitis (Non-infectious): Includes neurotrophic keratitis (due to corneal nerve damage), interstitial keratitis (immune-mediated), phlyctenular keratoconjunctivitis (hypersensitivity reaction).
* Peripheral Ulcerative Keratitis: Often associated with systemic autoimmune diseases.

3. Corneal Dystrophies

* Epithelial Basement Membrane Dystrophy (EBMD): Also called Map-Dot-Fingerprint dystrophy; causes recurrent erosions and irregular epithelium.
* Fuchs’ Endothelial Dystrophy: Progressive endothelial cell loss causing corneal edema and bullous keratopathy.
* Lattice Corneal Dystrophy: Amyloid deposits in stroma causing opacities and recurrent erosions.
* Schnyder’s Dystrophy: Crystalline lipid deposits in cornea with recurrent erosions.
* Granular and Macular Dystrophies: Stromal opacities affecting vision.

4. Corneal Degenerations

* Pellucid Marginal Degeneration: Thinning of inferior cornea with characteristic topographic changes.
* Terrien’s Marginal Degeneration: Peripheral thinning with lipid deposits and neovascularization.
* Band Keratopathy: Calcium deposition in superficial cornea.
* Salzmann’s Nodular Degeneration: Elevated nodules on corneal surface often after chronic inflammation or trauma.

5. Corneal Edema and Bullous Keratopathy

* Result from endothelial dysfunction (e.g., post-cataract surgery, Fuchs’ dystrophy).
* Presents with corneal swelling, pain, and decreased vision.

6. Trauma and Degenerative Conditions

* Corneal Abrasion: Superficial epithelial loss due to trauma.
* Corneal Scars: Result of healed ulcers or injuries causing opacity and vision loss.
* Corneal Erosions: Recurrent breakdown of epithelium, often linked to dystrophies or trauma.

7. Depositional and Metabolic Disorders

* Pseudoexfoliation Syndrome: Deposition of fibrillar material on anterior segment structures, including cornea.
* Bietti Crystalline Dystrophy: Rare lipid crystal deposits affecting cornea and retina.

**EPIDEMIOLOGY**

Eye trauma accounts for about 3% of all emergency department visits, with approximately 80% of these visits for corneal abrasions or foreign bodies. The incidence of corneal abrasion is higher among people of working age, with automotive workers between the ages of 20 and 29 years having the highest incidence of eye injuries.

Ocular chemical burns are an emergency, accounting for 11.5%–22.1% of all ocular injuries. Haring et al found that young children in the U.S. were the highest-risk individual group for ocular chemical burns, with ages 1 to 2 years at greatest risk. Generally, ocular chemical injuries occurred in individuals aged 18 to 64 years of age, making up the second most common cause of workplace ocular injuries after foreign bodies in the eye. Unfortunately, in recent years assault and hate crimes have contributed to an increased incidence of chemical injury, including up to 33% of severe ocular burns injuries.

**References**

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[Corneal Ulcer: Symptoms, Causes & Treatment](https://my.clevelandclinic.org/health/diseases/22524-corneal-ulcer)

**CYTOMEGALOVIRUS (CMV) RETINITIS**

**Definition and description**

Cytomegalovirus (CMV) retinitis is a serious eye infection. It often happens in people with immune systems that don’t work as well as they should. The term “retinitis” means inflammation of your retina, which is the part of your eye that senses light.

CMV is a necrotizing virus, meaning that the virus destroys tissue. It’s part of the herpes family of viruses. Most people get CMV at some point in their lives and don’t even know it. But the virus can reactivate in people with weakened (compromised) systems, sometimes causing CMV retinitis.

This includes people who:

* Have HIV/AIDS.
* Are receiving chemotherapy.
* Take immunosuppressant medications because they’ve had an organ transplant, bone marrow transplant or have autoimmune conditions.

CMV retinitis can result in long-term vision loss. In fact, it’s the main cause of blindness among people with HIV/AIDS.

### **Causes cytomegalovirus (CMV) retinitis**

If you’re infected with CMV, the virus can affect your retina, causing CMV retinitis. In adults with weakened immune systems, this is usually a reactivation of the virus rather than a new virus.

### **Signs and symptoms of cytomegalovirus retinitis**

You may not have symptoms of CMV retinitis right away. When you do develop them, symptoms may include:

* Floaters in your eye.
* Blind spots that move (scintillating scotoma).
* Decreased side vision (peripheral vision).
* Seeing wavy images (metamorphopsia).
* Blurred vision.
* Sensitivity to light (photophobia).
* Red eye.
* Eye pain.

The symptoms may start in one eye and then go on to your other eye.

**Diagnosis methods**

An ophthalmologist, or another type of eye care provider, will talk to you and ask you about your medical history and symptoms. Then they’ll do an eye examination after dilating your eyes.

Dilated pupils open widely, making it easier to see inside your eyes. Your provider will examine your retinas and other areas of your eyes for signs of disease. If you have retinitis, your provider may order lab work to identify its cause.

## **Management and Treatment**

Antiviral medicines can treat CMV retinitis, but they can’t cure it.

Your provider may decide to give you one — or a combination — of the following antiviral medications:

* Valganciclovir, given orally.
* Ganciclovir, given intravenously (IV) or as an intravitreal injection (IVI, or a shot into your eye).
* Foscarnet, given as an IV or IVI.
* Cidofovir, given as an IV or IVI.
* Letermovir, given orally.
* Fomiversen.

Treatment takes weeks because there’s an initial period of therapy followed by maintenance therapy.

## **Outlook / Prognosis**

The best prognosis (outlook) happens with prompt treatment. Untreated CMV retinitis can lead to low vision, even to the point of blindness.

Another condition that may happen as a result of CMV retinitis is retinal detachment, which means that your retina tears away from its supporting tissue. You can have surgery to repair this serious condition.

## **Living With**

If you have any sudden changes in eyesight — like dark spots in your vision or pain — get immediate medical help. Retinal detachment is a medical emergency.

It’s important to have regular eye exams if you have a weakened immune system.

### **What questions should I ask my healthcare provider about CMV retinitis?**

You may want to ask your provider some questions about CMV retinitis, including:

* How often should I schedule appointments?
* Is there anything I should avoid?
* How long will treatment last?
* When should I call you or go to an emergency room?
* Can you provide resources to help me cope with vision loss?
* Am I eligible for clinical trials?

#### **Complications of CMV retinitis treatment**

Antiviral treatment of CMV retinitis may cause your immune system to have a strong response, which can lead to uveitis. Uveitis causes eyes to be red, swollen and painful.

Sometimes, the drugs that treat CMV retinitis stop working for you. There are treatments that may help with ganciclovir-resistant CMV retinitis. These include leflunomide and T-lymphocyte infusion.

## **Differential diagnosis**

Early CMV can resemble cotton wool spots. It may be confused with HIV retinopathy, which is actually more common than CMV retinitis. HIV retinopathy occurs in 50-70% of patients and is characterized by intraretinal hemorrhages, cotton wool spots, and microaneurysms. For small lesions that are hard to differentiate from cotton wool spots, serial exams should be performed, which will show enlargement of the CMV lesions. Immunosuppression should be treated like a spectrum; some patients are more immunosuppressed than other individuals. For cases of minor immunosuppression, CMV retinitis may present as acute retinal necrosis (ARN) syndrome but follows a chronic course similar to CMV retinitis, which may cause misdiagnosis.

Syphilitic hemorrhagic necrotizing retinitis can mimic CMV retinitis; all patients should undergo treponemal testing.

**Epidemiology**

The incidence of IRU varies and is influenced by factors such as the HAART era and immune status at the time of initiation. Early reports estimated that IRU occurred in 10-17% of CMV retinitis (CMVR) cases during the initial HAART era, with over half of the cases developing inflammation after only a modest increase in CD4+ T-cell count (100–150 cells/mm³).

The timing of HAART initiation in CMVR is critical. Starting HAART before completing CMV induction therapy increases the risk of IRU. Conversely, controlling CMVR prior to immune reconstitution significantly reduces both the occurrence and severity of IRU. Ongoing anti-CMV therapy is advised to suppress viral antigen load until adequate immune recovery is achieved.

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**Diabetic retinopathy**

**Definition and description**

Diabetic Retinopathy occurs when there is damage to the blood vessels of the retina caused by diabetes, leading to progressive vision loss and also one of the leading causes of blindness in American adults. Regular eye exams are crucial for early detection and treatment .

Diabetic retinopathy (die-uh-BET-ik ret-ih-NOP-uh-thee) is a diabetes complication that affects eyes. It's caused by damage to the blood vessels of the light-sensitive tissue at the back of the eye (retina).

At first, diabetic retinopathy might cause no symptoms or only mild vision problems. But it can lead to blindness.

The condition can develop in anyone who has type 1 or type 2 diabetes. The longer you have diabetes and the less controlled your blood sugar is, the more likely you are to develop this eye complication.

**CAUSES**

Over time, too much sugar in your blood can lead to the blockage of the tiny blood vessels that nourish the retina, cutting off its blood supply. As a result, the eye attempts to grow new blood vessels. But these new blood vessels don't develop properly and can leak easily.

There are two types of diabetic retinopathy:

* **Early diabetic retinopathy.** In this more common form — called nonproliferative diabetic retinopathy (NPDR) — new blood vessels aren't growing (proliferating).  
  When you have NPDR, the walls of the blood vessels in your retina weaken. Tiny bulges protrude from the walls of the smaller vessels, sometimes leaking fluid and blood into the retina. Larger retinal vessels can begin to dilate and become irregular in diameter as well. NPDR can progress from mild to severe as more blood vessels become blocked.  
  Sometimes retinal blood vessel damage leads to a buildup of fluid (edema) in the center portion (macula) of the retina. If macular edema decreases vision, treatment is required to prevent permanent vision loss.
* **Advanced diabetic retinopathy.** Diabetic retinopathy can progress to this more severe type, known as proliferative diabetic retinopathy. In this type, damaged blood vessels close off, causing the growth of new, abnormal blood vessels in the retina. These new blood vessels are fragile and can leak into the clear, jellylike substance that fills the center of your eye (vitreous).  
  Eventually, scar tissue from the growth of new blood vessels can cause the retina to detach from the back of your eye. If the new blood vessels interfere with the normal flow of fluid out of the eye, pressure can build in the eyeball. This buildup can damage the nerve that carries images from your eye to your brain (optic nerve), resulting in glaucoma.

**RISK FACTOR**

Anyone who has diabetes can develop diabetic retinopathy. The risk of developing the eye condition can increase as a result of:

* Having diabetes for a long time
* Poor control of your blood sugar level
* High blood pressure
* High cholesterol
* Pregnancy
* Tobacco use
* Being Black, Hispanic or Native American

**SYMPTOMS**

You might not have symptoms in the early stages of diabetic retinopathy. As the condition progresses, you might develop:

* Spots or dark strings floating in your vision (floaters)
* Blurred vision
* Fluctuating vision
* Dark or empty areas in your vision
* Vision loss

**DIAGNOSIS**

Diabetic retinopathy is best diagnosed with a comprehensive dilated eye exam. For this exam, drops placed in your eyes widen (dilate) your pupils to allow your doctor a better view inside your eyes. The drops can cause your close vision to blur until they wear off, several hours later.

During the exam, your eye doctor will look for abnormalities in the inside and outside parts of your eyes.

Fluorescein angiography

After your eyes are dilated, a dye is injected into a vein in your arm. Then pictures are taken as the dye circulates through your eyes' blood vessels. The images can pinpoint blood vessels that are closed, broken or leaking.

Optical coherence tomography (OCT)

With this test, pictures provide cross-sectional images of the retina that show the thickness of the retina. This will help determine how much fluid, if any, has leaked into retinal tissue. Later, OCT exams can be used to monitor how treatment is working.

**TREATMENT**

Treatment, which depends largely on the type of diabetic retinopathy you have and how severe it is, is geared to slowing or stopping the progression.

Early diabetic retinopathy

If you have mild or moderate nonproliferative diabetic retinopathy, you might not need treatment right away. However, your eye doctor will closely monitor your eyes to determine when you might need treatment.

Work with your diabetes doctor (endocrinologist) to determine if there are ways to improve your diabetes management. When diabetic retinopathy is mild or moderate, good blood sugar control can usually slow the progression.

Advanced diabetic retinopathy

If you have proliferative diabetic retinopathy or macular edema, you'll need prompt treatment. Depending on the specific problems with your retina, options might include:

* **Injecting medications into the eye.** These medications, called vascular endothelial growth factor inhibitors, are injected into the vitreous of the eye. They help stop growth of new blood vessels and decrease fluid buildup.  
  Three drugs are approved by the U.S. Food and Drug Administration (FDA) for treatment of diabetic macular edema — faricimab-svoa (Vabysmo), ranibizumab (Lucentis) and aflibercept (Eylea). A fourth drug, bevacizumab (Avastin), can be used off-label for the treatment of diabetic macular edema.  
  These drugs are injected using topical anesthesia. The injections can cause mild discomfort, such as burning, tearing or pain, for 24 hours after the injection. Possible side effects include a buildup of pressure in the eye and infection.  
  These injections will need to be repeated. In some cases, the medication is used with photocoagulation.
* **Photocoagulation.** This laser treatment, also known as focal laser treatment, can stop or slow the leakage of blood and fluid in the eye. During the procedure, leaks from abnormal blood vessels are treated with laser burns.  
  Focal laser treatment is usually done in your doctor's office or eye clinic in a single session. If you had blurred vision from macular edema before surgery, the treatment might not return your vision to normal, but it's likely to reduce the chance of the macular edema worsening.
* **Panretinal photocoagulation.** This laser treatment, also known as scatter laser treatment, can shrink the abnormal blood vessels. During the procedure, the areas of the retina away from the macula are treated with scattered laser burns. The burns cause the abnormal new blood vessels to shrink and scar.  
  It's usually done in your doctor's office or eye clinic in two or more sessions. Your vision will be blurry for about a day after the procedure. Some loss of peripheral vision or night vision after the procedure is possible.
* **Vitrectomy.** This procedure uses a tiny incision in your eye to remove blood from the middle of the eye (vitreous) as well as scar tissue that's tugging on the retina. It's done in a surgery center or hospital using local or general anesthesia.

While treatment can slow or stop the progression of diabetic retinopathy, it's not a cure. Because diabetes is a lifelong condition, future retinal damage and vision loss are still possible.

Even after treatment for diabetic retinopathy, you'll need regular eye exams. At some point, you might need additional treatment.

**PREVENTION**

You can't always prevent diabetic retinopathy. However, regular eye exams, good control of your blood sugar and blood pressure, and early intervention for vision problems can help prevent severe vision loss.

If you have diabetes, reduce your risk of getting diabetic retinopathy by doing the following:

* **Manage your diabetes.** Make healthy eating and physical activity part of your daily routine. Try to get at least 150 minutes of moderate aerobic activity, such as walking, each week. Take oral diabetes medications or insulin as directed.
* **Monitor your blood sugar level.** You might need to check and record your blood sugar level several times a day — or more frequently if you're ill or under stress. Ask your doctor how often you need to test your blood sugar.
* **Ask your doctor about a glycosylated hemoglobin test.** The glycosylated hemoglobin test, or hemoglobin A1C test, reflects your average blood sugar level for the two- to three-month period before the test. For most people with diabetes, the A1C goal is to be under 7%.
* **Keep your blood pressure and cholesterol under control.** Eating healthy foods, exercising regularly and losing excess weight can help. Sometimes medication is needed, too.
* **If you smoke or use other types of tobacco, ask your doctor to help you quit.** Smoking increases your risk of various diabetes complications, including diabetic retinopathy.
* **Pay attention to vision changes.** Contact your eye doctor right away if your vision suddenly changes or becomes blurry, spotty or hazy.

Remember, diabetes doesn't necessarily lead to vision loss. Taking an active role in diabetes management can go a long way toward preventing complications.

**ALTERNATIVE MEDICINE**

Several alternative therapies have suggested some benefits for people with diabetic retinopathy, but more research is needed to understand whether these treatments are effective and safe.

Let your doctor know if you take herbs or supplements. They can interact with other medications or cause complications in surgery, such as excessive bleeding.

It's vital not to delay standard treatments to try unproven therapies. Early treatment is the best way to prevent vision loss.

**PROGNOSIS**

The prognosis of diabetic retinopathy depends on the duration of diabetes, glycemic control, associated comorbid conditions, and patient compliance to the appropriate line of treatment. Proper patient counseling is needed about their retinal condition and making patients aware that delay in proper follow-up could lead to permanent, irreversible vision loss. The initial stages of diabetic retinopathy are reversible if proper glycemic control is achieved. Many patients with diabetic macular edema require long-term support of repeated injections of intravitreal anti-VEGF medications. Patients treated with pan-retinal photocoagulation may require additional supplementation of anti-VEGF medications if there is persistent macular edema and neovascularization. Once there is tractional macular detachment for a longer duration, the visual prognosis is usually regarded as the macular anatomy is markedly distorted.

OCT biomarkers of prognosis of diabetic retinopathy include refractile bodies, disorganization of inner layers of the retina (DRIL), disruption of outer layers of the retina (DORL), choroidal thickness, epiretinal membrane, vitreomacular adhesions, subretinal fluid, macular thickness, and integrity of ellipsoid zone

**COMPLICATIONS**

Diabetic retinopathy involves the growth of abnormal blood vessels in the retina. Complications can lead to serious vision problems:

* **Vitreous hemorrhage.** The new blood vessels may bleed into the clear, jellylike substance that fills the center of your eye. If the amount of bleeding is small, you might see only a few dark spots (floaters). In more-severe cases, blood can fill the vitreous cavity and completely block your vision.  
  Vitreous hemorrhage by itself usually doesn't cause permanent vision loss. The blood often clears from the eye within a few weeks or months. Unless your retina is damaged, your vision will likely return to its previous clarity.
* **Retinal detachment.** The abnormal blood vessels associated with diabetic retinopathy stimulate the growth of scar tissue, which can pull the retina away from the back of the eye. This can cause spots floating in your vision, flashes of light or severe vision loss.
* **Glaucoma.** New blood vessels can grow in the front part of your eye (iris) and interfere with the normal flow of fluid out of the eye, causing pressure in the eye to build. This pressure can damage the nerve that carries images from your eye to your brain (optic nerve).
* **Blindness.** Diabetic retinopathy, macular edema, glaucoma or a combination of these conditions can lead to complete vision loss, especially if the conditions are poorly managed.

**When to see a doctor / red flag**

Careful management of your diabetes is the best way to prevent vision loss. If you have diabetes, see your eye doctor for a yearly eye exam with dilation — even if your vision seems fine.

Developing diabetes when pregnant (gestational diabetes) or having diabetes before becoming pregnant can increase your risk of diabetic retinopathy. If you're pregnant, your eye doctor might recommend additional eye exams throughout your pregnancy.

Contact your eye doctor right away if your vision changes suddenly or becomes blurry, spotty or hazy.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis of diabetic macular edema includes:

* Hypertensive retinopathy
* Central retinal vein occlusion
* Branch retinal vein occlusion
* Irvine Gass syndrome
* Post uveitic macular edema
* Ruptured microaneurysm
* Macular edema secondary to epiretinal membrane
* Choroidal neovascular membrane.

The diseases which can be mistaken as diabetic retinopathy based on the general fundus appearance include:

* Central retinal vein occlusion
* Hypertensive retinopathy
* Sickle cell retinopathy
* Terson syndrome
* Ocular ischemic syndrome
* Branch retinal vein occlusion
* Hemiretinal vein occlusion
* Valsalva retinopathy
* Post-traumatic retinal bleed
* Retinal macroaneurysm
* Retinopathy in thalassemia

**EPIDEMIOLOGY**

Diabetic retinopathy is one of the major neurovascular complications of diabetes and is a leading cause of blindness in adults of the working-age group. According to the recent epidemiological data shared by the American Academy of Ophthalmology, the global burden of diabetes mellitus is 387 million, which is estimated to increase to 592 million by 2035. Ninety-three million people are globally affected by diabetic retinopathy. Prevalence of diabetic retinopathy is 77.3% in type 1 diabetes patients and 25.1% in type 2 diabetes patients, out of which approximately 25% to 30% are expected to develop vision-threatening diabetic macular edema. Between 5% and 8% of patients with diabetic retinopathy need laser treatment. As many as 5% of patients will require vitrectomy surgery.

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**Dry eye syndrome**

**Definition and description**

Dry Eye Syndrome is a condition where there is insufficient tear production or poor tear quality causing eye discomfort and vision issues.

Dry eye disease is a common condition that occurs when your tears aren't able to provide adequate lubrication for your eyes. Tears can be inadequate and unstable for many reasons. For example, dry eyes may occur if you don't produce enough tears or if you produce poor-quality tears. This tear instability leads to inflammation and damage of the eye's surface.

Dry eyes feel uncomfortable. If you have dry eyes, your eyes may sting or burn. You may experience dry eyes in certain situations, such as on an airplane, in an air-conditioned room, while riding a bike or after looking at a computer screen for a few hours.

Treatments for dry eyes may make you more comfortable. These treatments can include lifestyle changes and eye drops. You'll likely need to take these measures indefinitely to control the symptoms of dry eyes.

**Causes and risk factors**

Dry eyes are caused by a variety of reasons that disrupt the healthy tear film. Your tear film has three layers: fatty oils, aqueous fluid and mucus. This combination usually keeps the surface of your eyes lubricated, smooth and clear. Problems with any of these layers can cause dry eyes.

Reasons for tear film dysfunction are many, including hormone changes, autoimmune disease, inflamed eyelid glands or allergic eye disease. For some people, the cause of dry eyes is decreased tear production or increased tear evaporation.

Decreased tear production

Dry eyes can occur when you're unable to produce enough liquid tears, also called aqueous fluid. The medical term for this condition is keratoconjunctivitis sicca (ker-uh-toe-kun-junk-tih-VY-tis SIK-uh). Common causes of decreased tear production include:

Aging

Certain medical conditions including Sjogren's syndrome, allergic eye disease, rheumatoid arthritis, lupus, scleroderma, graft vs. host disease, sarcoidosis, thyroid disorders or vitamin A deficiency

Certain medicines, including antihistamines, decongestants, hormone replacement therapy, antidepressants, and medicines for high blood pressure, acne, birth control and Parkinson's disease

Corneal nerve desensitivity caused by contact lens use, nerve damage or laser eye surgery, though symptoms of dry eyes related to this procedure are usually temporary

Increased tear evaporation

The oil film produced by small glands on the edge of your eyelids (meibomian glands) might become clogged. Blocked meibomian glands are more common in people with rosacea or other skin disorders.

Common causes of increased tear evaporation include:

Posterior blepharitis (meibomian gland dysfunction)

Blinking less often, which tends to occur with certain conditions, such as Parkinson's disease; or when you're concentrating during certain activities, such as while reading, driving or working at a computer

Eyelid problems, such as the lids turning outward (ectropion) and the lids turning inward (entropion)

Eye allergies

Preservatives in topical eye drops

Wind, smoke or dry air

Vitamin A deficiency

**RISK FACTORS**

Factors that make it more likely that you'll experience dry eyes include:

* Being older than 50. Tear production tends to diminish as you get older. Dry eyes are more common in people over 50.
* Being a woman. A lack of tears is more common in women, especially if they experience hormonal changes due to pregnancy, using birth control pills or menopause.
* Eating a diet that is low in vitamin A, which is found in liver, carrots and broccoli, or low in omega-3 fatty acids, which are found in fish, walnuts and vegetable oils.
* Wearing contact lenses or having a history of refractive surgery.

**Signs and symptoms**

Signs and symptoms, which usually affect both eyes, may include:

* A stinging, burning or scratchy sensation in your eyes
* Stringy mucus in or around your eyes
* Sensitivity to light
* Eye redness
* A sensation of having something in your eyes
* Difficulty wearing contact lenses
* Difficulty with nighttime driving
* Watery eyes, which is the body's response to the irritation of dry eyes
* Blurred vision or eye fatigue

**Diagnosis methods**

Tests and procedures that may be used to determine the cause of your dry eyes include:

* **A comprehensive eye exam.** An eye exam that includes a complete history of your overall health and your eye health can help your eye care specialist diagnose the cause of your dry eyes.
* **A test to measure the volume of your tears.** Your eye care specialist may measure your tear production using the Schirmer tear test. In this test, blotting strips of paper are placed under your lower eyelids. After five minutes your eye care specialist measures the amount of strip soaked by your tears.  
  Another option for measuring tear volume is the phenol red thread test. In this test, a thread filled with pH-sensitive dye (tears change the dye color) is placed over the lower eyelid, wetted with tears for 15 seconds and then measured for tear volume.
* **A test to determine the quality of your tears.** Other tests use special dyes in eye drops to determine the surface condition of your eyes. Your eye care specialist looks for staining patterns on the corneas and measures how long it takes before your tears evaporate.
* **A tear osmolarity test.** This type of test measures the composition of particles and water in your tears. With dry eye disease, there will be less water in your eyes.
* **Tear samples to look for markers** of dry eye disease, including elevated matrix metalloproteinase-9 or decreased lactoferrin.

**Treatment options**

For most people with occasional or mild dry eye symptoms, it's enough to regularly use nonprescription eye drops, also called artificial tears. If your symptoms are persistent and more serious, you have other options. What you do depends on what's causing your dry eyes.

Some treatments focus on reversing or managing a condition or factor that's causing your dry eyes. Other treatments can improve your tear quality or stop your tears from quickly draining away from your eyes.

### **Treating the underlying cause of dry eyes**

In some cases, treating an underlying health issue can help clear up the signs and symptoms of dry eyes. For instance, if a medication is causing your dry eyes, your eye care specialist may recommend a different medicine that doesn't cause that side effect.

If you have an eyelid condition, such as your lids turned outwards (ectropion), your eye care specialist may refer you to an eye surgeon who specializes in plastic surgery of the eyelids (oculoplastic surgeon).

### **Medications**

Prescription medicines used to treat dry eyes include:

* **Medicines to reduce eyelid inflammation.** Inflammation along the edge of your eyelids can keep oil glands from secreting oil into your tears. Your eye care specialist may recommend antibiotics to reduce inflammation. Antibiotics for dry eyes are usually taken by mouth, though some are used as eye drops or ointments.
* **Eye drops to control cornea inflammation.** Inflammation on the surface of your eyes (cornea) may be controlled with prescription eye drops that contain the immune-suppressing medicine cyclosporine (Restasis) or corticosteroids. Corticosteroids are not ideal for long-term use due to possible side effects.
* **Eye inserts that work like artificial tears.** If you have moderate to severe dry eye symptoms and artificial tears don't help, another option may be a tiny eye insert that looks like a clear grain of rice. Once a day, you place the hydroxypropyl cellulose (Lacrisert) insert between your lower eyelid and your eyeball. The insert dissolves slowly, releasing a substance that's used in eye drops to lubricate your eye.
* **Tear-stimulating medicines.** Medicines called cholinergics (pilocarpine, cevimeline) help increase tear production. These medicines are available as pills, gels or eye drops. Possible side effects include sweating.
* **Eye drops made from your own blood.** These are called autologous blood serum drops. They may be an option if you have severe dry eye symptoms that don't respond to any other treatment. To make these eye drops, a sample of your blood is processed to remove the red blood cells and then mixed with a salt solution.
* **A nasal spray to increase tear production.** The Food and Drug Administration (FDA) recently approved varenicline (Tyrvaya) to treat dry eyes. This medicine is delivered via a nasal spray. Varenicline is to be sprayed once into each nostril, twice a day.

### **Other procedures**

Other procedures that may be used to treat dry eyes include:

* **Closing your tear ducts to reduce tear loss.** Your eye care specialist may suggest this treatment to keep your tears from leaving your eye too quickly. This can be done by partially or completely closing your tear ducts, which normally serve to drain tears away.  
  Tear ducts can be plugged with tiny silicone plugs (punctal plugs). These are removable. Or tear ducts can be plugged with a procedure that uses heat. This is a more permanent solution called thermal cautery.
* **Using special contact lenses.** Ask your eye care specialist about newer contact lenses designed to help people with dry eyes.  
  Some people with severe dry eyes may opt for special contact lenses that protect the surface of the eyes and trap moisture. These are called scleral lenses or bandage lenses.
* **Unblocking oil glands.** Warm compresses or eye masks used daily can help clear up blocked oil glands. A thermal pulsation device is another way to unclog the oil glands, but it is unclear whether this method provides any advantage over warm compresses.
* **Using light therapy and eyelid massage.** A technique called intense-pulsed light therapy followed by massage of the eyelids may help people with severe dry eyes.

**Prevention tips**

If you experience dry eyes, pay attention to the situations that are most likely to cause your symptoms. Then find ways to avoid those situations in order to prevent your dry eyes symptoms. For instance:

* **Avoid air blowing in your eyes.** Don't direct hair dryers, car heaters, air conditioners or fans toward your eyes.
* **Add moisture to the air.** In winter, a humidifier can add moisture to dry indoor air.
* **Consider wearing wraparound sunglasses or other protective eyewear.** Safety shields can be added to the tops and sides of eyeglasses to block wind and dry air. Ask about shields where you buy your eyeglasses.
* **Take eye breaks during long tasks.** If you're reading or doing another task that requires visual concentration, take periodic eye breaks. Close your eyes for a few minutes. Or blink repeatedly for a few seconds to help spread your tears evenly over your eyes.
* **Be aware of your environment.** The air at high altitudes, in desert areas and in airplanes can be extremely dry. When spending time in such an environment, it may be helpful to frequently close your eyes for a few minutes at a time to minimize evaporation of your tears.
* **Position your computer screen below eye level.** If your computer screen is above eye level, you'll open your eyes wider to view the screen. Position your computer screen below eye level so that you won't open your eyes as wide. This may help slow the evaporation of your tears between eye blinks.
* **Stop smoking and avoid smoke.** If you smoke, ask your health care provider for help devising a quit-smoking strategy that's most likely to work for you. If you don't smoke, stay away from people who do. Smoke can worsen dry eye symptoms.
* **Use artificial tears regularly.** If you have chronic dry eyes, use eye drops even when your eyes feel fine to keep them well lubricated.

## 

## **Self care**

You may be able to manage your dry eyes with frequent eyelid washing and use of nonprescription eye drops or other products that help lubricate your eyes. If your condition is long term (chronic), use eye drops even when your eyes feel fine to keep them well lubricated.

### **Selecting and using nonprescription products for dry eyes**

A variety of nonprescription products for dry eyes are available, including eye drops, also called artificial tears, gels and ointments. Talk with your eye care specialist about which might be best for you.

Artificial tears may be all you need to control mild dry eye symptoms. Some people need to put drops in several times a day, and some use them only once a day.

Consider these factors when selecting a nonprescription product:

* **Preservative vs. non preservative drops.** Preservatives are added to some eye drops to prolong shelf life. You can use eye drops with preservatives up to four times a day. But using the preservative drops more often can cause eye irritation.  
  Non Preservative eye drops come in packages that contain multiple single-use vials. After you use a vial, you throw it away. If you rely on eye drops more than four times a day, non preservative drops are safe.
* **Drops vs. ointments.** Lubricating eye ointments coat your eyes, providing longer lasting relief from dry eyes. But these products are thicker than eye drops and can cloud your vision. For this reason, ointments are best used just before bedtime. Eye drops can be used at any time and won't interfere with your vision.
* **Drops that reduce redness.** It's best to avoid these as your solution for dry eyes, as prolonged use can cause irritation.

### **Washing your eyelids to control inflammation**

For people with blepharitis and other conditions that cause eyelid inflammation that blocks the flow of oil to the eye, frequent and gentle eyelid washing may help. To wash your eyelids:

* **Apply a warm washcloth to your eyes.** Wet a clean cloth with warm water. Hold the cloth over your eyes for five minutes. Rewet the cloth with warm water when it cools. Gently rub the washcloth over your eyelids — including the base of the eyelashes — to loosen any debris.
* **Use a mild soap on your eyelids.** Use baby shampoo or another mild soap. Put the cleanser on your clean fingertips and gently massage your closed eyes near the base of your eyelashes. Rinse completely.

## 

## **Alternative medicine**

Further study is needed, but some alternative medicine approaches may help relieve your dry eye symptoms. Discuss the benefits and risks with your eye care specialist.

* **Fatty acids.** Adding omega-3 fatty acids to your diet may help relieve dry eye signs and symptoms. These are available as supplements and in foods such as flaxseed, salmon and sardines.
* **Castor oil eye drops.** These eye drops may improve symptoms by reducing tear evaporation.
* **Acupuncture.** Some people have seen their dry eye symptoms improve after acupuncture therapy.

**Prognosis**

Dry eye is a chronic condition. It doesn’t have a cure, but treatments can help manage your symptoms.

You may have to try several different treatments to find what works best for you. This can be a stressful process, but it’s worth the effort. Talk to your provider if your current treatments aren’t working or you want to discuss other options.

**Possible complications**

People who have dry eyes may experience these complications:

* **Eye infections.** Your tears protect the surface of your eyes from infection. Without adequate tears, you may have an increased risk of eye infection.
* **Damage to the surface of your eyes.** If left untreated, severe dry eyes may lead to eye inflammation, abrasion of the corneal surface, corneal ulcers and vision loss.
* **Decreased quality of life.** Dry eyes can make it difficult to perform everyday activities, such as reading.

**When to see a doctor / red flag**

See your health care provider if you've had prolonged signs and symptoms of dry eyes, including red, irritated, tired or painful eyes. Your provider can take steps to determine what's bothering your eyes or refer you to a specialist.

**Differential diagnosis**

Many conditions may evoke symptoms similar to those caused by DED.Some conditions may also be associated with or lead to DED, such as allergic conjunctivitis, cicatricial conjunctivitis, filamentary keratitis, and neurotrophic keratitis. Identifying the underlying primary condition in these cases is key to reducing the progression of the disease and worsening of dry eye.

Differential diagnosis for DED include:

* Conjunctivitis (allergic, viral, bacterial, parasitic/chlamydial)
* Anterior blepharitis
* Demodex blepharitis
* Cicatricial conjunctivitis (Stevens-Johnson Syndrome, mucous membrane pemphigoid)
* Bullous keratopathy
* Contact lens–related keratoconjunctivitis
* Eyelid malposition (entropion, ectropion) or abnormality (trichiasis) leading to ocular surface disease
* Keratitis (interstitial, filamentary, contact lens–related, neurotrophic)

**Epidemiology data**

Dry eye is more common in women than men (due to female hormonal effects on the lacrimal and Meibomian glands and ocular surface) and has an increased prevalence with age. Studies have shown that female gender is a risk factor in developing dry eye, with a prevalence that ranges from 12% to 22%.The prevalence of DED worldwide varies depending on the diagnostic criteria employed, which ranges from approximately 5% to 50% in population-based studies.

DES has been shown to be as high as 70% in visual terminal users. In general, it is more common in Black individuals and Asian populations when compared with White individuals, although geographic, climatic, and environmental variations may also be significant factors. Evaporative dry eye is considered the most common subtype of DED. There may be discordance between dry eye signs and symptoms, with signs being more prevalent and variable than symptoms.

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**DUANE SYNDROME**

*ALTERNATIVE NAMES:*

Yes, there are several names for Duane syndrome. They include:

* Duane retraction syndrome.
* Eye retraction syndrome.
* Retraction syndrome or congenital retraction syndrome.
* Duane radial ray syndrome.
* Stilling-Turk-Duane syndrome.

**DEFINITION / DESCRIPTION**

Duane syndrome is a type of strabismus, an eye misalignment that’s congenital (something you’re born with). If you have Duane syndrome, you’ll have difficulties moving your eyes from side to side. You may have trouble with one or both eyes looking inward toward your nose or outward toward your ear.

You may also have trouble looking upward or downward. Along with your eyes crossing, or being out of alignment with each other, your eyelids might move when you try to move your eyes. This is because the nerves that control the muscles in your eyes don’t work correctly.

About 80% to 90% of Duane syndrome cases are unilateral, which means they affect only one eye. The left eye is more commonly affected.

**TYPES OF DUANE’S SYNDROME**

Yes, there are different types of Duane syndrome: type 1, type 2 and type 3.

**Type 1 Duane syndrome**

Duane syndrome type 1 is the most common form. It accounts for about 78% of all cases.

If you have type 1, you’ll have trouble moving your eye outward toward your ear. Your eye will be able to cross inward toward your nose, but your eye opening typically becomes smaller and the eyeball retracts. Your eye may look crossed in toward your nose (esotropia).

**Type 2 Duane syndrome**

Duane syndrome type 2 is the least common form. It accounts for about 7% of people with the condition.

If you have type 2, you have a limited ability to move your eyes inward toward your nose. You will be able to turn your eye outward toward your ear but your eye opening typically becomes smaller and the eyeball retracts. Your eye may look turned out toward your ear (exotropia).

**Type 3 Duane syndrome**

Duane syndrome type 3 accounts for about 15% of total cases.

If you have type 3, your ability to move your eyes in either direction is limited. Also, your eye opening may become smaller and your eyeball retracts with certain gazes.

It’s estimated that 4% of the population of the U.S., or some 13 million people, have strabismus. It’s further estimated that about 1% to 5% of the total number of people with strabismus have Duane syndrome.

Duane syndrome is slightly more common in females.

**CAUSES**

Duane syndrome is genetic, but not typically inherited. The movement issues are due to problems with your cranial nerves, or the nerves that control the way your eyes move.

Duane syndrome may happen more frequently in people who’ve been exposed to thalidomide in utero.

Only about 10% of people with Duane syndrome have other family members with the condition. In cases that run in the family, Duane syndrome is usually bilateral (it affects both eyes).

If you inherit Duane syndrome, it happens because a variant of *CHN1, MAFB,* or *SALL4* is passed down. The pattern for inheritance in Duane syndrome, called “autosomal dominant,” means that it occurs equally regardless of sex, and it can be inherited if just one parent has Duane syndrome.

**Is Duane syndrome contagious?**

No, Duane syndrome isn’t contagious. You can’t pass it on to someone like you can with an infection.

**SIGNS / SYMPTOMS**

Signs and symptoms of Duane syndrome, some of which may be seen in infants, may include:

* An unusual position of the head. People with Duane syndrome sometimes turn their heads to the side to straighten their eyes.
* Eyes that don’t point in the same direction, either at some times or all times (strabismus).
* One eye — the one that’s affected — having lower vision (amblyopia, or lazy eye).
* One eye seems smaller than the other because the eyelids narrow.
* Eyes that move either up or down when you’re looking a certain way. This is called an up-shoot or down-shoot.

**Do people with Duane syndrome have other conditions?**

Duane syndrome doesn’t always happen with other conditions, but it sometimes does. These conditions may include:

* Hearing disorders.
* Goldenhar syndrome.
* Problems with your spine or vertebrae.

There may also be other eye-related conditions that happen along with Duane syndrome, including:

* Cataract; which refers to the clouding of the lens of your eye.
* Microphthalmos; which means that your eye is unusually small.
* Nystagmus; a condition in which your eyes move uncontrollably.
* Crocodile tears syndrome, where someone with Bell’s palsy sheds tears while eating or drinking.

**DIAGNOSIS METHOD**

Your healthcare provider will take a complete medical history and will do a thorough eye exam. They’ll measure how much misalignment is present and test how far your eyes, or your child’s eyes, can move from side to side.

Your provider may also want to order tests for other conditions that might be linked to Duane syndrome.

There’s another condition called sixth cranial nerve palsy that’s very similar to Duane syndrome but is much less common than Duane syndrome.

## **Management and Treatment**

Treatment will depend on age, severity and other illnesses you might have.

Some people with Duane syndrome won’t need treatment. They’ll just need to keep a regular appointment schedule with their eye care providers.

People who also have amblyopia may benefit from patching their good eye so the weaker one becomes stronger.

If you have a severe case of Duane syndrome, you may need surgery on your eye muscles. The surgery can’t fix the nerve problem but it can change the position of eye muscles to relieve an abnormal head posture, straighten eyes when looking straight ahead, and improve up-shoots/down-shoots.

**PREVENTION TIPS**

At this time, there’s no way to prevent Duane syndrome from happening. Thalidomide, the medication that can cause Duane syndrome (in addition to many other congenital conditions), is no longer available.

**OUTLOOK / PROGNOSIS**

Some people with Duane syndrome can manage the condition by moving their heads slightly to improve their vision. In some cases, you may need surgery.

If you have another condition along with Duane syndrome, your provider will treat that condition also.

You may need glasses to improve vision and/or alignment.

## **Living With**

You may want to ask your healthcare provider questions like these:

* Should I see a genetic counselor?
* Are there any aids that may help with vision?

**DIFFERENTIAL DIAGNOSIS**

1. Abducens Nerve (Sixth Cranial Nerve) Palsy
   * Usually acquired (trauma, infection, neoplasm, vascular).
   * Absence of globe retraction and palpebral fissure narrowing on adduction.
   * Typically no upshoot or downshoot on adduction.
   * May have associated systemic neurological signs.
2. Moebius Syndrome
   * Congenital palsy of the facial (VII) and abducens (VI) nerves.
   * Bilateral facial weakness and inability to abduct the eyes.
   * No globe retraction or fissure narrowing.
3. Brown Syndrome
   * Limitation of elevation in adduction due to superior oblique tendon sheath abnormalities.
   * No limitation of abduction or globe retraction.
4. Congenital Fibrosis of Extraocular Muscles (CFEOM)
   * Congenital restrictive ophthalmoplegia involving multiple extraocular muscles.
   * Ptosis common; no globe retraction.
5. Marcus–Gunn Jaw Winking Syndrome
   * Synkinetic eyelid movement linked to jaw motion.
   * Ptosis with abnormal eyelid movements, no horizontal gaze limitation.
6. Congenital Esotropia
   * Large-angle esotropia presenting in infancy.
   * No globe retraction or fissure narrowing.
7. Okihiro Syndrome (Duane-Radial Ray Syndrome)
   * DRS with radial ray defects (thumb/hands).
   * Autosomal dominant with *SALL4* mutations.
8. Goldenhar Syndrome (Oculo-Auriculo-Vertebral Spectrum)
   * Facial asymmetry, ear anomalies, vertebral defects.
   * May have DRS-like eye movement abnormalities.
9. Wildervanck Syndrome
   * Triad of Klippel-Feil anomaly (cervical vertebrae fusion), hearing loss, and DRS.
10. Holt–Oram Syndrome
    * Upper limb and cardiac abnormalities with possible DRS.
11. Morning Glory Syndrome
    * Congenital optic disc anomaly; may be associated with DRS.
12. Other Congenital Cranial Dysinnervation Disorders (CCDDs)
    * Includes isolated horizontal gaze palsy without globe retraction, syndromes with scoliosis (HGPPS), and Bosley-Salih-Alorainy syndrome.

**Epidemiology**

Duane retraction syndrome (DRS) prevalence is estimated at between 1/1000 and 1/10,000 in the general population, representing approximately 1-5% of all strabismus cases. Females are more frequently affected than males.

REFERENCE:

[**Duane Retraction Syndrome - EyeWiki**](https://eyewiki.org/Duane_Retraction_Syndrome)

[**Duane Syndrome: Types, Causes & Management**](https://my.clevelandclinic.org/health/diseases/24315-duane-syndrome)

**ECTROPION**

**Definition and description**

Ectropion (ek-TROH-pee-on) is a condition in which your eyelid turns outward. This leaves the inner eyelid surface exposed and prone to irritation.

Ectropion is more common in older adults, and it generally affects only the lower eyelid. In severe ectropion, the entire length of the eyelid is turned out. In less severe ectropion, only one segment of the eyelid sags away from the eye.

Artificial tears and lubricating ointments can help relieve symptoms of ectropion. But usually, surgery is needed to fully correct the condition.

Ectropion can be caused by:

· **Muscle weakness.** As you age, the muscles under your eyes tend to weaken, and tendons stretch out. These muscles and tendons hold your eyelid taut against your eye. When they weaken, your eyelid can begin to droop.

· **Facial paralysis.** Certain conditions, such as Bell's palsy, and certain types of tumors can paralyze facial nerves and muscles. Facial paralysis that affects eyelid muscles can lead to ectropion.

· **Scars or previous surgeries.** Skin that has been damaged by burns or trauma, such as a dog bite, can affect the way that your eyelid rests against your eye. Previous eyelid surgery (blepharoplasty) can cause ectropion, particularly if a considerable amount of skin was removed from the eyelid at the time of surgery.

· **Eyelid growths.** Benign or cancerous growths on your eyelid can cause the lid to turn outward.

· **Genetic disorders.** Rarely is ectropion present at birth (congenital). When it is, it's usually associated with genetic disorders, such as Down syndrome.

**Risk factors**

Factors that increase your risk of developing ectropion include:

· **Age.** The most common cause of ectropion is weakening muscle tissue associated with aging.

· **Previous eye surgeries.** People who have had eyelid surgery are at higher risk of developing ectropion later.

· **Previous cancer burns or trauma.** If you've had spots of skin cancer on your face, facial burns or trauma, you're at higher risk of developing ectropion.

**Signs and symptoms**

Normally when you blink, your eyelids distribute tears evenly across your eyes, keeping the surfaces of the eyes lubricated. These tears drain into small openings on the inner part of your eyelids (puncta).

If you have ectropion, your lower lid pulls away from your eye and tears don't drain properly into the puncta. The resulting signs and symptoms can include:

· **Watery eyes (excessive tearing).** Without proper drainage, your tears may pool and constantly flow over your eyelids.

· **Excessive dryness.** Ectropion can cause your eyes to feel dry, gritty and sandy.

· **Irritation.** Stagnant tears or dryness can irritate your eyes, causing a burning sensation and redness in your eyelids and the whites of your eyes.

· **Sensitivity to light.** Stagnant tears or dry eyes can irritate the surface of the cornea, making you sensitive to light.

## **Diagnosis**

Ectropion can usually be diagnosed with a routine eye exam and physical. Your doctor may pull on your eyelids during the exam or ask you to close your eyes forcefully. This helps him or her assess each eyelid's muscle tone and tightness.

If your ectropion is caused by a scar, tumor, previous surgery or radiation, your doctor will examine the surrounding tissue as well.

Understanding how other conditions cause ectropion is important in choosing the correct treatment or surgical technique.

## **Treatment**

If your ectropion is mild, your doctor might recommend artificial tears and ointments to ease the symptoms. Surgery is generally required to fully correct ectropion.

### **Surgery**

The type of surgery you have depends on the condition of the tissue surrounding your eyelid and on the cause of your ectropion:

· **Ectropion caused by muscle and ligament relaxation due to aging.** Your surgeon will likely remove a small part of your lower eyelid at the outer edge. When the lid is stitched back together, the tendons and muscles of the lid will be tightened, causing the lid to rest properly on the eye. This procedure is generally relatively simple.

· **Ectropion caused by scar tissue from injury or previous surgery.** Your surgeon might need to use a skin graft, taken from your upper eyelid or behind your ear, to help support the lower lid. If you have facial paralysis or significant scarring, you might need a second procedure to completely correct your ectropion.

Before surgery, you'll receive a local anesthetic to numb your eyelid and the area around it. You may be lightly sedated using oral or intravenous medication to make you more comfortable, depending on the type of procedure you're having and whether it's done in an outpatient surgical clinic.

After surgery you might need to:

· Wear an eye patch for 24 hours

· Use an antibiotic and steroid ointment on your eye several times a day for one week

· Use cold compresses periodically to decrease bruising and swelling

After surgery you will likely experience:

· Temporary swelling

· Bruising on and around your eye

Your eyelid might feel tight after surgery. But as you heal, it will become more comfortable. Stitches are usually removed about a week after surgery. You can expect the swelling and bruising to fade in about two weeks.

**Lifestyle and home remedies**

These lifestyle tips may relieve your discomfort from ectropion:

· **Use eye lubricants.** Artificial tears and eye ointments can help keep your cornea lubricated and prevent vision-threatening damage. Using an eye ointment and wearing a moisture shield over your eye is particularly useful overnight.

· **Wipe your eyes carefully.** Constantly wiping watery eyes can make your under-eye muscles and tendons stretch even further, worsening your ectropion. Wipe from the outer eye up and in toward the nose.

**What you can do**

Before your appointment take these steps:

· List symptoms you've been having and for how long.

· Find a photo of yourself before the appearance of your eyelid that you can bring to the appointment.

· List all medications, vitamins and supplements you take, including the doses.

· List key personal and medical information, including other conditions, recent life changes and stressors.

· List questions to ask your doctor.

· Ask a relative or friend to accompany you, to help you remember what the doctor says.

For ectropion, some basic questions to ask your doctor include:

· What's the most likely cause of my symptoms?

· What kinds of tests do I need? Do they require any special preparation?

· Is this condition temporary or long lasting?

· Can ectropion damage my vision?

· What treatments are available, and which do you recommend?

· What are the risks of surgery?

· What are the alternatives to surgery?

· I have these other health conditions. How can I best manage them together?

· Do you have any brochures or other printed material that I can take with me? What websites do you recommend?

### **What to expect from your doctor**

Your doctor is likely to ask you a number of questions, such as:

· When did you begin experiencing symptoms?

· Have your symptoms been continuous or occasional?

· Have you had any previous surgery or procedures on your eye or eyelid?

· Have you had any radiation treatments on your head and neck?

· Have you had any other eye problems, such as an eye infection or an injury?

· Are you taking any blood thinners?

· Are you taking aspirin?

· Are you using any eye drops?

**Prevention**

Because entropion often occurs naturally with aging or after scarring, it’s difficult to prevent. To reduce your risk of developing entropion caused by injury, wear protective eyewear during activities that could injure your eye.

**Outlook / Prognosis**

Most people who receive treatment for entropion before it causes eye damage have a good outcome. Entropion surgery usually resolves the problem, and the condition rarely returns.

It’s important to treat entropion to avoid complications that may become permanent. Complications associated with an inward-turning eyelid include:

· Eye infections

· Corneal abrasions (scratches)

· Vision loss

**Complications**

Ectropion leaves your cornea irritated and exposed, making it more susceptible to drying. The result can be abrasions and ulcers on the cornea, which can threaten your vision.

### **When to see a doctor**

See your doctor if your eyes are constantly watering or irritated, or your eyelid seems to be sagging or drooping.

Seek immediate care if you have been diagnosed with ectropion and you experience:

· Rapidly increasing redness in your eyes

· Sensitivity to light

· Decreasing vision

These are signs and symptoms of cornea exposure or ulcers, which can harm your vision.

**DIFFERENTIAL diagnosis**

Entropion should be differentiated from epiblepharon, trichiasis, trachoma, and distichiasis.

1. Eyelid Retraction
   * Seen in thyroid eye disease (Graves’ orbitopathy) causing eyelid elevation and scleral show.
   * Unlike ectropion, the eyelid margin is not everted but abnormally elevated.
   * May follow lower eyelid surgery (blepharoplasty) or inferior rectus muscle recession without proper retractor disinsertion.
2. Floppy Eyelid Syndrome
   * Characterized by easily everted, lax upper eyelids, often associated with obesity and sleep apnea.
   * Upper eyelid eversion rather than lower eyelid ectropion.
   * Eyelid laxity is extreme but eyelid margin position differs from ectropion.
3. Eyelid Malignancy
   * Tumors (basal cell carcinoma, squamous cell carcinoma) can cause mechanical ectropion by distorting eyelid anatomy.
   * May present with ulceration, madarosis (loss of lashes), or infiltration.
4. Facial Nerve Palsy (Paralytic Ectropion)
   * Paralysis of orbicularis oculi muscle leads to lower eyelid laxity and ectropion.
   * Associated with other facial weakness signs.
5. Cicatricial Eyelid Changes
   * Scarring from burns, trauma, chronic inflammation, or surgery causes anterior lamellar shortening leading to ectropion.
   * Eyelid margin is pulled outward and may be fixed.
6. Mechanical Ectropion
   * Caused by mass effect from lesions such as chalazion, dermatochalasis, or orbital tumors pushing the eyelid margin outward.
7. Congenital Ectropion
   * Rare, usually bilateral, associated with syndromes such as blepharophimosis or euryblepharon.

**Epidemiology**

The older an individual is, the greater the chances of developing an entropion. Bilateral disease is three times more common than unilateral. Entropion is thought to occur more frequently in women than men, as women tend to have smaller tarsal plates than men

* Prevalence and Age Distribution  
  Ectropion, particularly involutional (age-related) ectropion of the lower eyelid, is common in the elderly population. Its prevalence increases markedly with age:
  + About 1% of people aged 60–69 years have ectropion.
  + This rises to approximately 7% in those aged 70–79 years.
  + In individuals over 80 years, prevalence may reach 17% to 49% depending on the study population.
  + One large study reported involutional ectropion prevalence of 49% in patients over 80 years, 33% in those 70–79, and 14% in 60–69 years. Only 5% of patients under 60 had ectropion.
* Gender Differences
  + Ectropion is more common in men than women.
  + Prevalence reported as high as 5.1% in men versus 1.5% in women in one elderly cohort.
  + Another large study found about 58% of ectropion patients were male.
* Associated Conditions and Risk Factors
  + Chronic periocular inflammatory diseases such as blepharitis, chalazion, hordeolum, eyelid dermatitis, and chronic conjunctivitis are strongly associated with involutional ectropion.
  + Systemic comorbidities including hypertension, dyslipidemia, and autoimmune rheumatic diseases also increase ectropion risk.
  + Age-related changes in eyelid tissues (collagen, elastin degeneration) and increased eyelid laxity contribute to pathogenesis.
  + Environmental factors like ultraviolet (UV) exposure, especially in rural areas, may increase risk.
* Prevalence Compared to Entropion
  + Ectropion is generally more prevalent than involutional entropion in elderly populations (e.g., 2.9% vs. 2.1% in one large study).
  + Entropion tends to be more common in women, while ectropion predominates in men.
* Other Forms
  + Cicatricial ectropion caused by scarring is less common but significant, with incidence varying widely depending on underlying conditions (45–80% in some studies of cicatricial eyelid disease).
  + Congenital ectropion is rare but reported in isolated case series, mostly bilateral and seen in neonates

REFERENCES

[Ectropion - Diagnosis and treatment - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/ectropion/diagnosis-treatment/drc-20351169)

[Ectropion - Symptoms and causes - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/ectropion/symptoms-causes/syc-20351164)

**ENDOPHTHALMITIS**

**Definition and description**

Endophthalmitis (pronounced en-dof-thal-**my-tis**) is the medical name for an infection that affects the inside of your eye. In endophthalmitis, a bacterial or fungal infection triggers inflammation, an immune system response.

Endophthalmitis affects the aqueous humor and the vitreous humor. The aqueous humor is a normally clear fluid found between your lens and cornea at the front of your eye. The vitreous humor, a clear gel, sits between your lens and retina toward the back of your eye.

Endophthalmitis is a serious medical condition that needs immediate treatment. It can spread and cause vision loss. **Panophthalmitis** is the name for an infection that spreads to all parts of your eyeball and extends into the orbit.

An ophthalmologist may say that you have bacterial endophthalmitis or fungal endophthalmitis. They may also name the type of endophthalmitis by indicating how you got the infection. For example:

· **Exogenous endophthalmitis**: This happens when something gets in your eye. You may develop exogenous endophthalmitis if you have an eye injury.

· **Endogenous endophthalmitis**: You may develop this type of endophthalmitis because you have a fungal or bacterial infection in another part of your body that then spreads to the eye through your blood. Endogenous endophthalmitis is less common than exogenous endophthalmitis.

### **What causes endophthalmitis?**

There are many causes of endophthalmitis. For example, any eye trauma, such as a tree branch hitting your eye or having eye surgery, may lead to exogenous endophthalmitis. Likewise, many infections in the body could lead to endogenous endophthalmitis, but this is usually in people who are severely ill.

#### **What are examples of penetrating eye trauma and exogenous endophthalmitis?**

Common causes of ways that bacteria and fungi can get into your eye from the outside include:

· **Eye surgery**: The most common eye surgery is cataract surgery, but you can also have surgery for glaucoma and other eye problems. Most people who develop postoperative endophthalmitis usually notice decreased vision and eye aches within a week or so after the procedure. Acute endophthalmitis comes on quickly after surgery. Chronic endophthalmitis takes longer to develop and may last longer.

· **Injections into the eye**, called intraocular or intravitreal injections: Eye care providers may treat some condition by injecting certain medications into your vitreous humor. Age-related macular degeneration is example of a condition treated with intraocular or intravitreal injections.

· **Industrial or motor vehicle accidents**: You can get a foreign object that penetrates your eye from an industrial or motor vehicle accident.

· **Sports**: You can hurt your eye while playing contact sports.

#### **What kinds of bacterial and fungal infections cause endogenous endophthalmitis?**

In Europe and North America, the most common bacteria that cause endogenous endophthalmitis are *Staphylococcus aureus* and *Streptococcus pneumoniae*. *Candida albicans* causes a majority of fungal infections*.*

Bacterial and fungal infections can spread through your body by getting into your bloodstream. Here are a few examples of these infections:

· Having dental work.

· Receiving intravenous drugs (through the vein, or IV).

· Having pneumonia or sepsis.

· Having surgical procedures anywhere inside your body.

· Having an abscess or a skin ulcer.

· Having a urinary tract infection.

Recent studies have found that some people who’ve had COVID-19 developed endogenous endophthalmitis. It’s most likely that the endophthalmitis resulted from an infection that developed in the hospital. The people most at risk seem to be people who:

· Were in the hospital for a long time.

· Have other serious medical conditions.

· Were treated with IV antibiotics and other medicines.

##### **Is endophthalmitis contagious?**

Although infectious agents cause endophthalmitis, it doesn’t spread from person to person.

**Signs and symptoms**

Endophthalmitis signs and symptoms may include:

· Red eyes.

· Significant eye pain.

· Loss of vision.

· Sensitivity to bright light (photophobia).

· Swollen eyelids.

· Watery eyes (epiphora).

**Diagnosis methods**

An eye care provider will ask you about your health history and your current signs and symptoms. They’ll give you a complete eye exam.

They’ll take a sample of fluid or discharge from your eye to test in a laboratory. They may also want to test samples of your blood and urine.

In some cases, a provider might ask for an ultrasound of the eye (ocular ultrasound).

## **Management and Treatment**

An eye care provider may treat endophthalmitis with medicine. They may prescribe antibiotic or antifungal medications or corticosteroids. You may get eye drops or injections.

If the disease severely affects your vision, your provider may suggest a surgery called a vitrectomy. This procedure removes infectious material from your eye and lets the surgeon inject antibiotic or antifungal medication inside your eye.

Your provider may suggest you wear an eye patch for a time after the surgery.

## **Prevention**

Ways to reduce your risk of endophthalmitis may include:

1. Always using protective eye gear if your job involves work where an object like a piece of machinery or a tool might hit your eye. You should also wear protective eye gear if you play contact sports.

2. Making sure your hands are clean if you have to touch your eyes.

3. Following provider recommendations for self-care after surgery.

## **Outlook / Prognosis**

As with many conditions, the outlook for endophthalmitis is typically better if it’s caught and treated early. Outcomes may be worse in people who have immune systems that don’t work well or have diabetes.

Some people have low vision that lasts after treatment. If this happens, speak to your healthcare team about getting support.

## **Living With**

If you have endophthalmitis, you’ll need regular eye appointments. Always tell a healthcare provider about any change in vision or eye pain.

If you have vision loss after treatment, your provider can help you find support services if they’re necessary.

#### **Complications/side effects of the treatment**

Possible complications from intravitreal injections of anti-infective drugs include damage to the cornea or the retina.

· Panophthalmitis

· Glaucoma

· Orbital cellulitis

· Intracranial spread

· Septicemia

· Loss of vision

· Phthisis

· Hypotony

· Painful blind eye

**DIFFERENTIAL DIAGNOSIS**

**Post-surgical Endophthalmitis**

1. Toxic anterior segment syndrome (TASS) is an acute inflammatory condition presenting within 24 hours of surgery with a severe anterior chamber reaction, limbus to limbus corneal edema. However, there is no adnexal involvement, and the patient responds dramatically to the addition of steroids. Various features differentiating TASS from acute postoperative endophthalmitis are listed in figure 4 (common differentiating features of TASS with endophthalmitis).

2. Posteriorly dislocated lens matter

3. Fibrinous reaction post vitrectomy

4. Phacoanaphylaxis etc.

**Non-surgical Endophthalmitis**

1. Chronic uveitis – certain cases of chronic indolent endophthalmitis may be confused for chronic panuveitis.

2. Retained intraocular foreign body

3. Old vitreous hemorrhage

4. Toxoplasma retinochoroiditis

5. Necrotic retinoblastoma

6. Acute retinal necrosis

7. Severe panuveitis

8. Seasonal hyperacute panuveitis (SHAPU)

9. Intravitreal cysticercus with intraocular inflammation

**EPIDEMIOLOGY**

The relative frequency of various subtypes of endophthalmitis varies depending upon the geography, level of specialization at the ophthalmic center, and study duration. For example, studies before 2005 have very few cases of post intravitreal anti-VEGF (vascular endothelial growth factor) injection endophthalmitis as anti-VEGF agents received first FDA approval in 2004.

In a recent retrospective cohort study in the U.S., acute onset endophthalmitis occurred in 0.04% of cataract surgeries and 0.016% of patients receiving an intravitreal injection. The incidence of post-traumatic endophthalmitis varies from 0.9% to 17%.

Case series from Asian countries have reported an incidence of 0.023% to 0.076% of endophthalmitis in post-cataract surgery setting and 0.01% to 0.10% post intravitreal injections.

Among the intraocular surgeries, secondary intraocular lens implantation seems to have a very high risk (0.36%) for acute endophthalmitis, and trabeculectomy may have a high risk (1.8%, maybe up to 5.7%) for delayed or late-onset endophthalmitis especially after the advent of mitomycin-C or 5-fluorouracil.

Endophthalmitis after pars plana vitrectomy had the lowest rates (0.046%) when compared with other ocular surgeries [secondary IOL-0.366%, combined penetrating keratoplasty (PK) with cataract surgery- 0.194%, PK- 0.178%, glaucoma surgeries -0.124%, combined trabeculectomy and cataract surgery- 0.114%, overall incidence after intraocular surgery-0.093%, and cataract surgery with or without IOL-0.082%] in a study.

There is no significant difference in the rates of endophthalmitis after intravitreal injection of various anti-VEGF agents. Cluster endophthalmitis cases have occurred after intravitreal use of bevacizumab due to either lapse in the cold chain, sub-optimal compounding of bevacizumab aliquots, or fake drug.

#### **How soon after treatment will I feel better?**

Your symptoms of pain and redness may begin to improve after a few days, but you may continue to have vision problems after that. It can take weeks or even months for endophthalmitis to resolve completely.

REFERENCES

[Endophthalmitis: Causes, Symptoms & Treatment](https://my.clevelandclinic.org/health/diseases/24468-endolphthalmitis)

<https://www.ncbi.nlm.nih.gov/books/NBK559079/>

**EYE MELANOMA**

**Definition and description**

Eye melanoma is a kind of eye cancer that starts in cells within the eye that make melanin. Melanin is most often known as the pigment that gives skin its color. But the eyes have cells that make melanin too. Eye melanoma also is called ocular melanoma, intraocular melanoma and uveal melanoma.

Most eye melanomas form in parts of the eye you can't see when looking in a mirror. That makes eye melanoma hard to notice. And eye melanoma typically doesn't cause symptoms at first.

Eye melanoma can be treated. Treatment for small eye melanomas may not cause vision problems. But treatment for large eye melanomas typically leads to some vision loss.

**Causes**

It's not clear what causes eye melanoma.

Eye melanoma happens when cells in the eye develop changes in their DNA. A cell's DNA holds the instructions that tell the cell what to do. In healthy cells, the DNA tells the cells to grow and multiply at a set rate. The DNA also tells the cells to die at a set time.

In cancer cells, the DNA changes give different instructions. The changes tell the cancer cells to grow and multiply quickly. Cancer cells can keep living when healthy cells die. This causes too many cells.

The cancer cells might form a mass called a tumor. The tumor can grow to invade and destroy healthy tissue. In time, cancer cells can break away and spread to other parts of the body. When cancer spreads, it's called metastatic cancer.

**Where eye melanoma develops**

Eye melanoma most often happens in the cells of the middle layer of the eye, called the uvea. The uvea has three parts. Each part can be affected by eye melanoma:

* **The iris.** The iris is the colored part at the front of the eye.
* **The choroid layer.** The choroid layer is a layer of blood vessels and connective tissue. It's located at the back of the uvea.
* **The ciliary body.** The ciliary body is located behind the iris. It helps the eye focus. It also makes a transparent liquid, called aqueous humor, that fills the front part of the eye.

Eye melanoma can happen in other parts of the eye. But this is very rare. Other parts of the eye that can develop melanoma include:

* The layer on the front of the eye, called the conjunctiva.
* The socket that surrounds the eyeball, called the orbit.
* The eyelid.

**Risk factors**

Risk factors for eye melanoma include:

* **Light eye color.** People with blue eyes or green eyes have a higher risk of melanoma of the eye.
* **Being white.** White people have a greater risk of eye melanoma than do people of other races.
* **Age.** The risk of eye melanoma goes up with age.
* **Certain inherited skin conditions.** A condition called dysplastic nevus syndrome, which causes unusual moles, can raise the risk of eye melanoma.

People who have a condition called ocular melanocytosis also are at higher risk of eye melanoma. This condition involves unusual skin pigmentation on the eyelids and in the tissue around the eyelids. It also leads to more pigmentation on the eye's uvea.

* **Certain genetic changes.** Some DNA changes that are passed from parents to children may raise the risk of eye melanoma.
* **Exposure to ultraviolet light.** Some research suggests that exposure to ultraviolet light could raise the risk of eye melanoma. Sources of ultraviolet light include the sun, as well as tanning beds.

Healthcare professionals haven't found anything that can prevent eye melanoma.

**Signs and symptoms**

Eye melanoma may not cause any symptoms. When they do happen, signs and symptoms of eye melanoma can include:

* Flashes of light or what look like specks of dust in a person's vision. These are sometimes called floaters.
* A growing dark spot in the colored part of the eye, called the iris.
* A change in the shape of the pupil. The pupil is the dark circle at the center of the eye.
* Poor vision or blurry vision in one eye.
* Not being able to see when looking to the side. This is called loss of peripheral vision.

**Diagnosis methods**

Eye melanoma diagnosis often starts with an eye exam. Imaging tests can help show the size of the cancer.

### **Eye exam**

During an eye exam for eye melanoma, a healthcare professional may first examine the outside of the eye. The health professional may look for blood vessels that are larger than usual. Large blood vessels might mean there's something concerning happening inside the eye.

An eye exam also involves looking inside the eye with the help of special equipment. One way to do that uses lenses and a bright light mounted on a healthcare professional's forehead. This is called binocular indirect ophthalmoscopy. Another method uses lenses and a microscope that has an intense beam of light that lights up the inside of the eye. This is called slit-lamp biomicroscopy.

### **Fundus photography**

Fundus photography is a test that takes color pictures of the inside surface of the eye. This part of the eye is called the fundus. Fundus photography can show an eye melanoma. The test might be repeated to watch a melanoma over time. Different kinds of tests can take pictures of the fundus to show an eye melanoma. One example is fundus autofluorescence.

### **Eye ultrasound**

An eye ultrasound uses high-frequency sound waves to make images of the eye. The sound waves come from a device that looks like a wand, called a transducer. A healthcare professional places the transducer on the closed eyelid or on the front surface of the eye to get the pictures.

### **Eye angiography**

Angiography is a test that makes pictures of the blood vessels. To get pictures of the blood vessels in the eye, a colored dye is injected into a vein in an arm. The dye travels to the blood vessels in the eye. A camera with special filters to detect the dye takes pictures of the eye every few seconds for several minutes. Tests that can make pictures of the blood vessels in the eye include fluorescein angiography and indocyanine green angiography.

### **Optical coherence tomography**

Optical coherence tomography is an imaging test that uses light waves to make pictures of the eye. It can make pictures of the uvea and the retina that might show an eye melanoma.

### **Eye melanoma biopsy**

A biopsy is a procedure to remove a sample of tissue for testing in a lab. A biopsy typically isn't necessary to diagnose eye melanoma. But it may be used in some situations. Sometimes a biopsy may be done at the time of treatment to get more information about the cancer cells.

### **Testing for cancer spread**

Other tests may be needed to see if melanoma has spread to other parts of the body. The tests may include:

* Blood tests.
* Tests to check how well the liver works.
* Chest X-ray.
* Ultrasound.
* Computerized tomography scan, also called CT scan.
* Magnetic resonance imaging, also called MRI.
* Positron emission tomography scan, also called PET scan.

**Treatment options**

**Treatment**

Not all eye melanomas need treatment. When treatment is needed, it can include radiation therapy, laser therapy, photodynamic therapy or surgery. Targeted therapy and immunotherapy may be used to treat eye melanoma in some situations.

Which treatment is best for eye melanoma depends on several factors. These factors include the size and location of the cancer. Treatment also depends on whether cancer has spread beyond the eye. Your overall health and what you prefer to do is part of treatment planning too.

**Waiting to treat small eye melanomas**

A small eye melanoma may not need to be treated right away. If the melanoma is small and isn't growing, you and your healthcare professional might choose to wait and watch for signs of growth.

If the melanoma grows or causes other health concerns, you may choose to have treatment at that time.

**Radiation therapy for eye melanoma**

Radiation therapy treats cancer with powerful energy. Radiation therapy is typically used for small to medium-sized eye melanomas.

For eye melanoma, radiation therapy often involves placing a radioactive device on the eye. The device is called a plaque. It looks like a bottle cap. The plaque holds several radioactive seeds. A healthcare professional places the plaque on the eye, over the cancer. The plaque is held in place with temporary stitches. The plaque stays in place for a few days. Then it's removed. Radiation treatment that involves putting the radiation inside the body is called brachytherapy.

The radiation also can come from a machine that aims beams of radiation, such as proton beams. The beams can be aimed at the eye to treat eye melanoma. Giving radiation with a machine outside the body is called external beam radiation. This type of radiation therapy often is given over several days.

**Laser therapy for eye melanoma**

Laser therapy uses a laser light to hurt the cancer cells. For eye melanoma, it might be used in some situations. One type of laser treatment, called thermotherapy, uses an infrared laser. It's sometimes used along with radiation therapy to treat eye melanoma.

**Photodynamic therapy for eye melanoma**

Photodynamic therapy is a two-stage treatment that combines light energy with a medicine called a photosensitizer. The photosensitizer kills cancerous and precancerous cells when activated by light, usually from a laser. For eye melanoma, photodynamic therapy is used for smaller cancers.

**Surgery for eye melanoma**

Surgery to treat eye melanoma may involve removing the melanoma or removing the entire eye.

* **Surgery to remove the melanoma.** Surgery to remove the melanoma and a band of healthy tissue around it may be an option for treating some small eye melanomas.
* **Surgery to remove the eye.** Surgery to remove an eye is called enucleation. It may be used for some large eye melanomas.

After the eye with melanoma is removed, an implant often is put into the same place. The muscles that control the movement of the eye are attached to the implant. That allows the implant to move. After the eye has time to heal, an artificial eye, called a prosthesis, is made that matches the other eye.

**Targeted therapy for eye melanoma**

Targeted therapy for cancer is a treatment that uses medicines that attack specific chemicals in the cancer cells. By blocking these chemicals, targeted treatments can cause cancer cells to die. For eye melanoma, targeted therapy may be used when the cancer has spread to other parts of the body or in situations where surgery isn't possible.

**Immunotherapy for eye melanoma**

Immunotherapy for cancer is a treatment with medicine that helps the body's immune system kill cancer cells. The immune system fights off diseases by attacking germs and other cells that shouldn't be in the body. Cancer cells survive by hiding from the immune system. Immunotherapy helps the immune system cells find and kill the cancer cells. For eye melanoma, immunotherapy may be used when the cancer has spread to other parts of the body or in situations where surgery isn't possible.

**Prevention tips**

The cause of ocular melanoma is largely unknown, and it isn’t clear if it can be prevented.

It’s not clear if there’s an association with sunlight exposure or how strong this association is. It’s possible that protecting your eyes from the sun may help reduce your risk of cancer.

The American Cancer Society recommends wearing UV-protected sunglasses when you’re outside in strong sunlight. Wrap-around glasses with at least 99 percent UVA and UVB protection are best.

**PROGNOSIS**

Local treatment prevents local recurrence in 95% of patients. However, due to micrometastasis, 50% of patients with uveal tumors develop metastatic disease. Metastatic disease is typically fatal, with a median survival of only 10 months after diagnosis. Several prognostic indicators leading to poor outcomes include increased patient age, the largest basal diameter of the tumor, ciliary body involvement, extrascleral tumor extension, epithelioid cell type, and vasculogenic mimicry patterns.

Chromosomal analysis identifying monosomy 3 and 8q gain can help determine prognosis to some extent, but gene expression profiling proves to have better prognostic abilities. Gene expression profiling assesses the expression of 12 discriminating genes and 3 control genes to determine the metastatic potential, classifying patients into either class 1, indicating low potential, or class 2, indicating high potential. In addition, detecting circulating tumor cells or DNA may be a prognostic indicator for metastatic disease.

The 5-year survival rate for conjunctival melanoma with treatment is 83% to 84%, and the 10-year survival rate is 69% to 80%. The overall mortality for choroidal melanoma and ciliary body melanoma is 30% to 50% within 10 years, primarily due to metastatic disease. The overall mortality rate for iris melanoma is 0% to 11%.

**Complications**

Some people develop other health concerns linked to eye melanoma. These are called complications. They can include the following:

**Vision loss**

Eye melanoma can cause vision loss. Sometimes vision loss is a symptom of an eye melanoma. Sometimes vision loss is caused by eye melanoma treatment.

**Eye melanoma that spreads beyond the eye**

Eye melanoma can spread outside of the eye to other areas of the body, including the liver, lungs and bones.

**When to see a doctor**

Make an appointment with a healthcare professional if you have any symptoms of eye melanoma. If you notice sudden changes in your vision, seek emergency medical care right away.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnoses for choroidal melanoma and ciliary body melanoma are as follows:

* Choroidal detachment
* Intraocular foreign body
* Chronic angle closure glaucoma
* Hyphema
* Cavernous hemangioma
* Vitreous hemorrhage
* Age-related macular degeneration
* Melanocytoma
* Medulloepithelioma
* Choroidal osteoma
* Adenoma
* Adenocarcinoma
* Combined hamartoma of the retina and pigment epithelium
* Congenital hypertrophy and reactive hyperplasia of the retinal pigment epithelium
* Lymphoid tumor
* Hemangiopericytoma
* Leiomyoma
* Neurofibroma
* Glioneuroma
* Astrocytoma
* Rhabdomyosarcoma
* Posterior uveitis
* Sarcoid nodules
* Tubercular granuloma
* Uveitis

The differential diagnoses for iris melanoma are as follows:

* Iris pigment epithelial cyst
* Iris stromal cyst
* Iris nevi
* Iris metastases
* Iris foreign body
* Iridocorneal endothelial syndrome
* Koeppe or Busacca nodules
* Lisch nodules
* Peripheral anterior synechiae
* Other iris tumors, such as leiomyoma and rhabdomyosarcoma
* Cogan-Reese syndrome

Clinicians must distinguish conjunctival melanoma from other conditions, including conjunctival squamous cell carcinoma; conjunctival melanosis; conjunctival mycosis; conjunctival seborrheic keratosis; acquired melanosis; foreign body like graphite; drug toxicity, such as epinephrine; and pseudomelanoma.

**EPIDEMIOLOGY**

Ocular melanoma is the most prevalent primary intraocular malignancy in adults, accounting for 3% to 5% of all melanomas. Uveal melanoma accounts for 85% to 90%, with 5% originating in the iris and the remainder arising from the ciliary body or choroid. The median age of diagnosis is approximately 62, with a peak incidence observed between 70 and 79.

White patients of northern European descent have the highest incidence of disease. In contrast, the incidence among Black patients is low, whereas the incidence in patients who are Asian or Hispanic is considered moderate. Men have a 30% higher incidence compared to females. In the United States, the incidence of ocular melanoma is approximately 5 per 1 million population. Internationally, in countries with large populations of individuals of northern European descent, the incidence is 7.5 per 1 million people annually.

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**Entropion**

**Definition and description**

Entropion is a condition that occurs when your eyelid turns inward toward your eyeball. When this happens, your eyelid skin and eyelashes rub against the surface of your eye. This friction often causes discomfort and irritation to your cornea (front of your eye). Entropion is the opposite of ectropion, the outward turning of your eyelid.

Entropion usually occurs in your lower eyelid, and it can happen on one side or both (bilateral entropion). Without treatment, it can damage your cornea and lead to vision loss. If you’ve developed the condition, it’s important to see an eye care specialist for treatment. Entropion treatment typically starts with noninvasive measures but may require surgery.

### **Types of entropion**

There are several types of entropion that can affect your eyes. These are:

* Involutional. Muscles and tendons in your eyes loosen and weaken as you age, resulting in this most common type of entropion.
* Spastic. Irritation, infection and inflammation can lead to this type. It most commonly occurs after eye surgery in people who had previously unrecognized involutional entropion.
* Cicatricial. Scar tissue formation causes this type of entropion. It may result from burns, infections, inflammation, trauma, eye surgery or radiation therapy.
* Congenital. This means you’ve had the condition since birth.
* Mechanical. The weight of your eyelid is causing it to turn in on itself. This type commonly occurs due to a mass or tumor on your eyelid.

### **Entropion causes**

There are three characteristics of an eyelid with entropion: eyelid looseness (laxity), eyelid retractor disinsertion and excessively strong eyelid closing muscles which cause your eyelid to turn in rather than out. Lower eyelid laxity is common in people over the age of 60 because their eyelid supports weaken with age. Other causes of an in-turned eyelid include:

* Eye injury
* Infection
* Previous eye surgery
* Inflammatory conditions
* Scarring, due to any of the above

### **Risk factors for entropion**

Aging is the biggest risk factor for entropion. It’s most common in people over 60 years old. According to the American Academy of Ophthalmology, it affects about 2.1% of people over the age of 60.

People with sleep apnea may be at greater risk of developing entropion, as they tend to have excessively loose upper eyelids (floppy eyelid syndrome), which are prone to turning in on themselves.

Entropion also affects women more often than men.

**Signs and symptoms**

### **Symptoms of entropion**

Entropion causes many uncomfortable symptoms in your eye, including:

* Feeling like there’s something in your eye
* Red eyes
* Tearing (watering)
* Blurry vision
* Eye pain
* Itching
* Sensitivity to light (photophobia) and wind
* Drainage or crusting

When you first develop entropion, you may only notice occasional symptoms. But over time, the symptoms usually become constant.

**Diagnosis methods**

Your eye care specialist will start by asking you about your symptoms and medical history. They’ll perform an eye exam, looking for any signs of skin irritation, infection or scarring. They’ll pay close attention to the edges of your eyelid, as there are other medical conditions that can mimic entropion. They want to ensure they make the correct diagnosis to decide the proper treatment.

#### **Tests that are used to diagnose entropion**

Your provider may perform several tests to help diagnose entropion. These tests include:

* Snap-back test. Without allowing you to blink, your provider pulls your eyelid down and observes how long it takes to return to its original position.
* Distraction test. Your provider pulls your eyelid away from your eye and measures the distance. They consider more than 6 millimeters abnormal.
* Slit lamp exam. A slit lamp is a special microscope with a bright light that your provider uses to examine your eye, looking for signs of the condition.

**Treatment options**

Your healthcare provider may treat entropion in several ways. Common entropion treatments include:

* Lubricating eye drops. Lubricating your eye with ointment and artificial tears can provide moisture and relieve discomfort. This isn’t a long-term solution, though.
* Soft contact lenses. Your healthcare provider might recommend wearing soft contact lenses to protect your eyes from irritation. Contact lenses act as a bandage to help ease your symptoms.
* Tape. You can use tape to reposition your eyelid temporarily.
* Stitches. Your provider will numb your eyelid and stitch it into a more favorable position. The resulting scar tissue can also help keep your eyelid turned outward for a few months.
* Botox®. Your provider can use a small amount of botulinum toxin to weaken the eyelid muscles that are causing your lid to turn in.
* Eyelash removal. Epilation is a procedure where your eyelashes are plucked to keep them from irritating the surface of your eye.

If your eyelid is turned in due to chronic inflammation, your provider may order labs or perform a biopsy to investigate rarer causes, like ocular cicatricial pemphigoid or trachoma.

#### **Entropion surgery**

The above options may offer a temporary fix, but your healthcare provider will usually have to perform surgery to address the inward-turning eyelid and return it to a normal position. These surgeries typically involve sedation and local anesthesia to numb your eyelid. You’ll go home the same day as your surgery.

Surgical entropion repair methods include:

* Eyelid tightening. This procedure shortens your eyelid (called a wedge resection or lateral tarsal strip) to tighten your lid.
* Retractor reinsertion. This procedure tightens your lid retractor (the muscle that opens and closes your lid).
* Orbicularis debulking. This procedure weakens part of the muscle that’s responsible for closing your eyelids.
* Eyelid margin reconstruction. For trauma or chronic inflammation, electrocautery and/or cryotherapy (freezing) may be used to permanently remove (epilate) misdirected eyelashes.

While nonsurgical treatments are helpful in the short term, entropion surgery is usually necessary to fully and permanently correct the condition.

Most people experience temporary side effects following entropion surgery, including swelling and bruising. These symptoms are a normal part of healing and you can manage them with prescribed medication and cold compresses. In most cases, people fully recover from surgical entropion repair in about two weeks.

**Lifestyle and home remedies**

To relieve the symptoms of entropion until you have surgery, you can try:

* **Eye lubricants.** Artificial tears and eye ointments help protect your cornea and keep it lubricated.
* **Skin tape.** Special transparent skin tape can be applied to your eyelid to keep it from turning in. Place one end of the tape near your lower eyelashes, then pull down gently and attach the other end of the tape to your upper cheek. Ask your doctor to demonstrate proper technique and placement of the tape.

**Prevention tips**

Because entropion often occurs naturally with aging or after scarring, it’s difficult to prevent. To reduce your risk of developing entropion caused by injury, wear protective eyewear during activities that could injure your eye

**Prognosis**

Most people who receive treatment for entropion before it causes eye damage have a good outcome. Entropion surgery usually resolves the problem, and the condition rarely returns.

It’s important to treat entropion to avoid complications that may become permanent. Complications associated with an inward-turning eyelid include:

* Eye infections
* Corneal abrasions (scratches)
* Vision loss

**Possible complications**

Corneal irritation and injury are the most serious complications related to entropion because they can lead to permanent vision loss.

**When to see a doctor / red flag**

Contact your healthcare provider if your eyelid turns inward. Even if your eyelid doesn’t appear to turn inward, seek medical treatment if you feel like something is constantly in your eye.

**Differential diagnosis**

Entropion should be differentiated from epiblepharon, trichiasis, trachoma, and distichiasis.

1. Epiblepharon
   * A horizontal fold of redundant skin and orbicularis muscle near the eyelid margin causes lashes to be directed vertically or slightly inward without true eyelid margin inversion.
   * Common in Asian children, often resolves with age.
   * Unlike entropion, the eyelid margin itself remains in normal position and lashes revert to normal when skin is pulled away.
2. Trichiasis
   * Misdirection of eyelashes toward the globe without eyelid margin malposition.
   * Can cause similar symptoms of irritation and corneal damage.
   * No inward turning of the eyelid margin.
3. Distichiasis
   * Presence of an extra row of eyelashes emerging from the meibomian gland orifices.
   * Lashes may be finer and cause irritation but eyelid margin is normal.
4. Ectropion
   * Outward turning of the eyelid margin, causing exposure and dryness rather than irritation from lashes.
   * Can cause epiphora and conjunctival redness.
5. Cicatricial Eyelid Changes
   * Scarring from trauma, burns, infections, or inflammation can cause cicatricial entropion or other lid malpositions.
   * Often associated with difficulty in lid eversion and skin changes.
6. Spastic Entropion
   * Caused by orbicularis muscle spasm leading to intermittent inward turning of the eyelid.
   * Can be distinguished by persistence of entropion after forced eyelid closure and specific clinical tests.

**Epidemiology data .**

The older an individual is, the greater the chances of developing an entropion. Bilateral disease is three times more common than unilateral. Entropion is thought to occur more frequently in women than men, as women tend to have smaller tarsal plates than men

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**Floaters**

**Definition and description**

Small spots or threads in vision caused by debris in the vitreous humor. Eye floaters are spots in your vision. They may look to you like black or gray specks, strings, or cobwebs. They may drift about when you move your eyes. Floaters appear to dart away when you try to look at them directly.

Most eye floaters are caused by age-related changes that occur as the jelly-like substance (vitreous) inside your eyes liquifies and contracts. Scattered clumps of collagen fibers form within the vitreous and can cast tiny shadows on your retina. The shadows you see are called floaters.

If you notice a sudden increase in eye floaters, contact an eye specialist immediately — especially if you also see light flashes or lose your vision. These can be symptoms of an emergency that requires prompt attention.

#### **What do eye floaters look like?**

There are many ways to describe eye floaters. Some people see spiders, amoebas or clouds. Your own creativity guides how you think a floater looks. Your description of floaters might sound completely different from someone else’s definition. If you have floaters, you might see:

* Squiggly lines.
* Spots.
* Spider Like shapes.
* Threadlike strands.
* Small shadowy shapes.
* Black or very dark spots.

In most cases, eye floaters are a normal and common part of the aging process. As you get older, the fluid within your eyes (vitreous) shrinks. This is normal and doesn’t mean that your eyes aren’t healthy.

It’s important to maintain regular eye exams over time, especially if you’re experiencing chronic floaters. Chronic floaters usually aren’t something you need to be concerned about, but it’s a good idea to have your eyes regularly checked to make sure there aren’t any other serious eye issues. If you experience an acute (sudden) onset of floaters and flashes, you should see an eye care provider.

#### **Can eye floaters happen in only one eye or both eyes at the same time?**

Your eyes may not age in the same way or at the exact same time. The vitreous might shrink in one eye a little faster than in the other. Often, you’ll have floaters in one eye at a time. It can happen to both of your eyes but usually not at the same time.

#### **Are there stages or different severity levels of eye floaters?**

Noticing eye floaters is normal, but if you notice a group of new floaters at one time, contact your eye care provider. Sudden changes in vision, including a number of new eye floaters, mean that you should set up an appointment with an eye specialist.

**CAUSES**

Eye floaters may be caused by vitreous changes related to aging or from other diseases or conditions:

## Age-related eye changes. The vitreous is a jelly-like substance made primarily of water, collagen (a type of protein) and hyaluronan (a type of carbohydrate). The vitreous fills the space in your eye between the lens and retina and helps the eye maintain its round shape. As you age, the vitreous changes. Over time, it liquifies and contracts — a process that causes it to pull away from the eyeball's inside surface. As the vitreous changes, collagen fibers within the vitreous form clumps and strings. These scattered pieces block some of the light passing through the eye. This casts tiny shadows on your retina that are seen as floaters.

## Inflammation in the back of the eye. Uveitis is inflammation in the middle layer of tissue in the eye wall (uvea). Posterior uveitis affects the back of the eye, which includes the retina and an eye layer called the choroid. The inflammation causes floaters in the vitreous. Causes of posterior uveitis include infection, autoimmune disorders and inflammatory diseases.

## Bleeding in the eye. Bleeding into the vitreous can have many causes, including retinal tears and detachments, diabetes, high blood pressure (hypertension), blocked blood vessels, and injury. Blood cells are seen as floaters.

## Torn retina. Retinal tears can happen when a contracting vitreous tugs on the retina with enough force to tear it. Without treatment, a retinal tear may lead to retinal detachment. If fluid leaks behind the tear, it can cause the retina to separate from the back of your eye. Untreated retinal detachment can cause permanent vision loss.

## Eye surgeries and eye medications. Certain medications that are injected into the vitreous can cause air bubbles to form. These bubbles are seen as shadows until your eye absorbs them. Silicone oil bubbles added during certain surgeries on the vitreous and retina also can be seen as floaters.

## **Risk factors**

## Factors that can increase your risk of eye floaters include:

## Age over 50 years

## Nearsightedness

## Eye injury

## Complications from cataract surgery

## Diabetes complication that causes damage to the blood vessels of the retina (diabetic retinopathy)

## Eye inflammation

## **Symptoms**

Symptoms of eye floaters may include:

* Small shapes in your vision that appear as dark specks or knobby, transparent strings of floating material
* Spots that move when you move your eyes, so when you try to look at them, they move quickly out of your line of vision
* Spots that are most noticeable when you look at a plain bright background, such as a blue sky or a white wall
* Small shapes or strings that eventually settle down and drift out of the line of vision

## **Diagnosis**

### Your eye care specialist conducts a complete eye exam to determine the cause of your eye floaters. Your exam usually includes eye dilation. Eye drops widen (dilate) the dark center of your eye. This allows your specialist to better see the back of your eyes and the vitreous.

### **Treatment**

### Most eye floaters don't require treatment. However, any medical condition that is the cause of eye floaters, such as bleeding from diabetes or inflammation, should be treated.

### Eye floaters can be frustrating and adjusting to them can take time. Once you know the floaters will not cause any more problems, over time you may be able to ignore them or notice them less often.

### If your eye floaters get in the way of your vision, which happens rarely, you and your eye care specialist may consider treatment. Options may include surgery to remove the vitreous or a laser to disrupt the floaters, although both procedures are rarely done.

### Surgery to remove the vitreous. An ophthalmologist who is a specialist in retina and vitreous surgery removes the vitreous through a small incision (vitrectomy). The vitreous is replaced with a solution to help your eye maintain its shape. Surgery may not remove all the floaters, and new floaters can develop after surgery. Risks of a vitrectomy include infection, bleeding and retinal tears.

### Using a laser to disrupt the floaters. An ophthalmologist aims a special laser at the floaters in the vitreous (vitreolysis). This may break up the floaters and make them less noticeable. Some people who have this treatment report improved vision; others notice little or no difference. Risks of laser therapy include damage to your retina if the laser is aimed incorrectly.

**Prevention tips**

You can’t prevent eye floaters that are due to age, but you may be able to reduce your risk of floaters by managing chronic illnesses like diabetes and high blood pressure (hypertension).

### **Are eye floaters an emergency?**

Eye floaters aren’t usually emergencies. You don’t need to worry if you see the occasional floater. You should let your eye care provider know about the floaters and have your eyes checked regularly to make sure there are no other vision issues.

However, if you suddenly have more floaters than normal, reach out to your healthcare provider right away. This could be a sign of a retinal tear or detachment. These conditions need immediate treatment.

**Possible complications**

There aren’t complications of floaters caused by aging. If floaters are due to retinal diseases, your vision can be at risk.

### **When to see a doctor**

Contact an eye specialist immediately if you notice:

* Many more eye floaters than usual
* A sudden onset of new floaters
* Flashes of light in the same eye as the floaters
* A gray curtain or blurry area that blocks part of your vision
* Darkness on a side or sides of your vision (peripheral vision loss)

These painless symptoms could be caused by a retinal tear, with or without a retinal detachment. This is a sight-threatening condition that requires immediate attention.

#### **What can I do at home to treat floaters?**

There are no home remedies for floaters.

**Differential Diagnosis**

The differential diagnosis includes:

* Vitreous floater/posterior vitreous detachment
* Vitreous hemorrhage
* Retinal tear or detachment

**EPIDEMIOLOGY**

A vitreous detachment typically affects patients older than 50 and increases in prevalence by age 80. Individuals who are myopic or nearsighted have an increased risk of vitreous floaters. Additionally, eyes with an inflammatory disease after direct trauma to the globe or having recently undergone eye surgery have an increased chance of developing a vitreous floater. Men and women appear to be affected equally.

* Prevalence:  
  Eye floaters are very common in the general population. Multiple studies report that approximately 74% to 76% of people experience floaters at some point. For example, a smartphone-based survey of 603 individuals worldwide found that 76% reported seeing floaters. Another similar survey reported 74% prevalence.
* Severity and Impact:  
  Among those who notice floaters, about 33% report moderate to severe visual impairment or bother from floaters. This means roughly one-third of people with floaters find them significantly disruptive.
* Age:  
  Although floaters are often associated with aging and posterior vitreous detachment (PVD), some studies found no significant difference in floater prevalence across age groups, including younger populations. However, clinical experience suggests floaters become more common and symptomatic with advancing age due to vitreous degeneration.
* Refractive Error Associations:
  + Myopia (nearsightedness): Myopes are about 3.5 times more likely to report moderate to severe floaters compared to emmetropes (normal vision).
  + Hyperopia (farsightedness): Hyperopes are even more likely, about 4.4 times more likely, to report moderate to severe floaters.  
    The exact mechanisms behind these associations remain unclear, but earlier onset of PVD in myopes may contribute.
* Other Factors:  
  No significant associations were found between floater prevalence and gender, race, or iris color. Comorbid eye conditions like diabetes, glaucoma, prior eye injury, or LASIK surgery showed trends toward increased floater severity but were not statistically significant in some studies.
* Global Distribution:  
  Floaters are reported worldwide with no significant variation by country or ethnicity in surveyed populations.

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**Fuchs' dystrophy**

**Definition and description**

Fuchs dystrophy is a condition in which fluid builds up in the clear tissue at the front of the eye, called the cornea. This causes your cornea to swell and thicken, leading to glare, blurred or cloudy vision, and eye discomfort.

Fuchs (fewks) dystrophy usually affects both eyes. It may cause your vision to get worse over time. The disease often starts in the 30s and 40s, but many people with Fuchs dystrophy don't develop symptoms until they reach their 50s or 60s.

Some medicines and self-care steps may help relieve symptoms of Fuchs dystrophy. When advanced disease causes more-serious vision problems, cornea transplant surgery is the best way to restore vision.

**Causes**

The cells lining the inside of the cornea are called endothelial cells. Those cells help maintain a healthy balance of fluid within the cornea and keep the cornea from swelling. In Fuchs dystrophy, the endothelial cells slowly die or do not work well, causing fluid buildup within the cornea. The fluid buildup, called edema, causes thickening of the cornea and blurred vision.

Fuchs dystrophy tends to run in families. The genetic basis of the disease is complex. Family members can be affected to different degrees or not at all.

**Risk factors**

Some factors make it more likely that you'll develop Fuchs dystrophy They include:

* **Sex.** Fuchs dystrophy is more common in women than in men.
* **Genetics.** Having a family history of Fuchs dystrophy increases your risk.
* **Age.** There is a rare early-onset type of Fuchs dystrophy that starts in childhood. Most cases start in the 30s and 40s, but many people with Fuchs dystrophy don't develop symptoms until their 50s or 60s.

**Signs and symptoms**

As Fuchs dystrophy gets worse, symptoms often affect both eyes. Symptoms may include:

* Blurred or cloudy vision, sometimes described as a lack of clear vision.
* Changes in vision throughout the day. Symptoms are worse in the morning when you wake up and slowly get better during the day. As the disease gets worse, blurred vision may take longer to get better or does not get better at all.
* Glare, which can decrease your vision in dim and bright light.
* Seeing halos around lights.
* Pain or grittiness from tiny blisters on the surface of your cornea.

**Diagnosis methods**

An eye care professional will test your vision. You also may have tests to help diagnose Fuchs dystrophy. Those tests may include:

* **Cornea examination and grading.** A member of your eye care team will use a special eye microscope called a slit lamp to look for drop-shaped bumps called guttae on the back surface of the cornea. This eye care professional will then check your cornea for swelling and stage your Fuchs dystrophy.
* **Corneal thickness.** An eye care professional may use a test called corneal pachymetry to measure the thickness of the cornea.
* **Corneal tomography.** Taking a special picture of your cornea helps an eye care professional look for swelling in your cornea. This test is called corneal tomography.
* **Corneal cell count.** Sometimes an eye care professional uses a special instrument to record the number, shape and size of the cells that line the back of the cornea. This test is not required.

**Treatment options**

Some nonsurgical treatments may help relieve symptoms of Fuchs dystrophy. If you have advanced disease, an eye care professional may suggest surgery.

### **Medicines and other therapies**

* **Eye medicine.** Saline (5% sodium chloride) eye drops or ointments can help reduce the amount of fluid in your cornea.
* **Soft contact lenses.** These act as a covering to relieve pain.

### **Surgery**

People who have surgery for advanced Fuchs dystrophy can have much better vision and remain symptom-free for years. Surgical options include:

* **Transplanting the inner layer of the cornea.** This is called Descemet membrane endothelial keratoplasty, also known as DMEK. In this procedure, the back layer of the cornea is replaced with healthy endothelial cells from a donor. It is usually done with local anesthesia in an outpatient setting.
* **Transplanting the cornea.** If you have another eye condition or already had eye surgery, DMEK may not be an option. An eye care professional may recommend a partial-thickness cornea transplant. This is called Descemet-stripping endothelial keratoplasty, also known as DSEK. In rare cases, a full-thickness cornea transplant may be done. This type of transplant is called penetrating keratoplasty, also known as PK.

### **Potential future treatments**

A variety of new treatments are being investigated that could change how Fuchs dystrophy is managed in the future. Since the discovery of the genetic mutation associated with most cases of Fuchs dystrophy, there is a better understanding of how the disease might develop. This offers the potential for nonsurgical therapies in the future. Various eye drop treatments are being developed and may enter clinical trials in the future. Novel surgical treatments also are being studied to find if they might be helpful.

**Lifestyle and home remedies**

Follow instructions from your eye care team to take care of your eyes. You also can try other things to help reduce glare and soothe your eyes.

* Use nonprescription salt solution (5% sodium chloride) eye drops or ointment.
* Dry your eyes with a hair dryer. Hold it at arm's length and direct warm — not hot — air across your face, especially in the morning when swelling is worse. This helps remove extra fluid in the cornea, which reduces swelling.

**Prevention tips**

There’s currently no way to prevent Fuchs’ corneal dystrophy. But you can help yourself by quitting smoking if you do smoke. If you have diabetes, work with your healthcare provider to manage your blood sugar levels.

**Prognosis**

Depending on how severe it is, Fuchs’ dystrophy may cause extreme pain, low vision or blindness if you don’t seek treatment.

With treatment, your expected outcome is much better. Your vision may improve to 20/20 within days of surgery (with glasses). That’s why it’s important to work with your provider to find the right treatment for you.

**When to see a doctor / red flag**

If you have some of these symptoms, and especially if they get worse over time, see an eye care professional. The eye care professional may refer you to a corneal specialist. If symptoms develop suddenly, call for an urgent appointment. Other eye conditions that cause the same symptoms as Fuchs dystrophy also require treatment right away.

## **Differential Diagnoses**

* Aphakic Bullous Keratopathy
* Interstitial Keratitis
* Keratopathy, Pseudophakic Bullous
* Other endothelial dystrophies such as posterior polymorphous dystrophy or congenital hereditary endothelial dystrophy (CHED)
* Postoperative Corneal Edema
* Recurrent Corneal Erosion
* Traumatic corneal decompensation

## **Epidemiology**

### Frequency

United States

Exact incidence of Fuchs endothelial dystrophy is not known. It begins with the formation of guttate excrescences. Cornea guttata is seen quite often. Frequency of cornea guttata increases with age. After age 40 years, 70% of patients have cornea guttata. Only 0.1% of these patients have epithelial edema and bullae formation.

International

A cross-sectional study in Japan found the prevalence of cornea guttata to be 4.1% among residents aged 40 years or older using only specular microscopic criteria.Older age, female sex, and a thinner cornea were independently associated with a higher risk of cornea guttata.

### Mortality/Morbidity

Once corneal decompensation starts, the course is relentless. In a matter of months or years, the vision is progressively disturbed. Finally, the patient is visually crippled. In addition, problems caused by repeated bullae formation, ulceration, scarring, and vascularization occur. If left untreated, the condition ends in near blindness, which may be painful.

### Race

No race is immune from this condition.

### Sex

Females are affected more than males (3:1).

### Age

Fuchs endothelial dystrophy can be differentiated into early-onset (manifesting in the third decade of life) and late-onset (manifesting in the sixth decade of life, on average). However, the root of the condition is evident 1 or 2 decades earlier in the form of profuse cornea guttata in the central part of the cornea.

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**Glaucoma**

**Definition and description**

Glaucoma is a group of diseases that damage the optic nerve, it is often associated with increased eye pressure, causing gradual vision loss and potentially blindness. It often progresses slowly without noticeable symptoms, earning it the nickname “sneak thief of sight”. It includes open-angle and angle-closure types.

Angle-closure glaucoma is one of two main types of glaucoma, a condition that’s defined by high pressure within your eye. The name for this pressure is intraocular pressure (IOP).

High IOP happens because fluid called aqueous humor can’t flow freely in your eye. In angle-closure glaucoma, a bulging or swollen iris (the colored part of your eye) blocks the drainage system. The condition may happen quickly or over time.

Other names for angle-closure glaucoma are closed-angle glaucoma and narrow-angle glaucoma.

**Types of angle-closure glaucoma**

There are different types of angle-closure glaucoma, including primary and secondary forms.

Primary angle-closure glaucoma

Acute angle-closure glaucoma comes on quickly and is a medical emergency because permanent vision damage can happen very quickly. Symptoms include severe eye pain, blurred vision, halos, nausea, vomiting and red eye.

Intermittent angle-closure glaucoma refers to cases where the drainage system changes from open to closed. You may or may not have symptoms.

Chronic angle-closure glaucoma may not cause symptoms at the beginning. But gradually, symptoms develop over time. Later, it might result in an episode of acute angle-closure glaucoma or a gradual increase of pressure with possible damage to your optic nerve.

Secondary angle-closure glaucoma

This form happens along with another condition. These include:

Poorly managed diabetes, which can cause a condition called proliferative diabetes-related retinopathy.

Poorly managed blood pressure or other vascular diseases that can lead to conditions like ischemic central vein occlusion.

Uveitis.

Membranes and scars from injury or inflammation.

Angle-closure glaucoma will happen to about 1 in 1,000 people. It’s most likely to happen after the age of 40.

One estimate is that 17.14 million people throughout the world who are over 40 have primary angle-closure glaucoma, with a majority (12.3 million people) in Asia.

**Symptoms and Causes**

Acute angle-closure glaucoma symptoms may include:

Severe eye pain.

* Redness.
* Vision loss.
* Seeing rainbow-colored rings around lights or halos.
* Headache.
* Nausea and vomiting.

When they’re present, which doesn’t always happen, symptoms of chronic angle-closure glaucoma and other forms of non-acute angle-closure glaucoma may include:

* Redness.
* Blurred vision.
* Eye discomfort.
* Headache or brow ache.

**What causes angle-closure glaucoma?**

Angle-closure glaucoma happens because aqueous humor in your eye can’t flow in and out as it normally should. Your body constantly makes new fluid to replace the existing fluid.

When the old fluid can’t leave your eye, it backs up and causes pressure. The pressure causes damage to your optic nerve.

There are many reasons that you can get glaucoma. With angle-closure glaucoma, the reason is that your natural lens thickens over time and pushes against your iris, blocking the drainage passages.

Research indicates that angle-closure glaucoma has an association with family history and certain genes.

**Risk factors for angle-closure glaucoma**

Risk factors for developing angle-closure glaucoma include:

Age: The risk rises with age as IOP increases and your eye changes.

Sex: The risk is higher for females.

Ethnicity: The risk is higher for people of Asian descent.

**What are the complications?**

Like other forms of glaucoma, angle-closure glaucoma can damage your optic nerve and cause permanent loss of vision.

**Diagnosis and Tests**

Any medical examination begins with your provider asking you questions about your symptoms and medical history, including information on your biological family’s history.

Your eye care provider will give you a thorough eye exam.

What tests will be done to diagnose this condition?

Your provider will diagnose angle-closure glaucoma using:

Gonioscopy. This test can assess the drainage angle in your eye.

Slit-lamp exam. A special microscope lets your provider see inside your eye.

Tests to measure eye pressure (IOP).

Testing your peripheral vision with visual field tests.

Imaging tests like optical coherence tomography or ultrasound.

If your provider thinks you have angle-closure glaucoma, they’ll avoid using drugs that dilate your pupils because those medications could make drainage worse.

**Management and Treatment**

How is angle-closure glaucoma treated?

There’s no cure for angle-closure glaucoma, but there are treatments.

Treatments for acute angle-closure glaucoma

Treatment for acute angle-closure glaucoma must happen quickly to avoid vision loss.

Your provider will offer treatments that may include:

Medications. These include timolol, pilocarpine and brimonidine eye drops and oral acetazolamide.

Laser iridotomy. This is a procedure that makes a hole in your iris to allow fluid to flow freely. If you have an episode of acute angle-closure glaucoma in one eye, your provider will do an iridotomy on both eyes. Otherwise, you’re at risk of having an episode in your other eye.

Treatment for non-acute angle-closure glaucoma

Your provider is likely to treat non-acute forms of angle-closure glaucoma with laser iridotomy.

Your provider may also suggest cataract removal, which should make the progression of chronic angle-closure glaucoma go more slowly.

**Complications/side effects of the treatments for angle-closure glaucoma**

Complications may include blurred vision and sensitivity to light for a few days. You may also experience a streak of light from the site of the laser treatment. You typically need a driver to take you home after the procedure, but you may start normal activities the next day.

**Prevention**

Can angle closure glaucoma be prevented?

No, you can’t prevent angle-closure glaucoma. But screening eye exams with an eye care provider will help you know if you have risks for angle closure glaucoma. They’ll then suggest treatment and medications to avoid if you’re at risk. It’s also very important to manage your blood sugar if you have diabetes.

**Outlook / Prognosis**

What can I expect if I have angle-closure glaucoma?

If you have treatment for angle-closure glaucoma, your outlook should be good. If you have acute angle-closure glaucoma, you need immediate treatment to preserve your vision.

You should listen to your provider’s suggestions on when and how often to return for eye exams.

**Living With**

When should I see my healthcare provider?

You should always see an eye care provider if you have changes in your vision. Follow the appointment schedule they suggest for you.

When should I go to an emergency room?

Times when you should get emergency medical help, like calling your eye care provider or going to an ER, include these events:

Symptoms that start up suddenly, like vision loss.

Severe eye pain.

A combination of symptoms, like red eye or nausea and eye pain.

An eye injury.

Symptoms that develop after any type of eye surgery.

What questions should I ask my doctor or eye care provider?

You may want to ask your healthcare team these questions:

What type of glaucoma do I have?

What treatment do you recommend?

Is the type of glaucoma I have hereditary?

What are the side effects of treatment?

What’s the outlook for my condition in the short term and the long term?

Are there activities that I should avoid?

How often should I schedule appointments?

**Causes and risk factors of glaucoma**

Glaucoma develops when the optic nerve becomes damaged. As this nerve gradually gets worse, blind spots develop in your vision. For reasons that eye doctors don't fully understand, this nerve damage is usually related to increased pressure in the eye.

Raised eye pressure happens as the result of a buildup of fluid that flows throughout the inside of the eye. This fluid, called the aqueous humor, usually drains through a tissue located at the angle where the iris and cornea meet. This tissue is called the trabecular meshwork. The cornea is important to vision because it lets light into the eye. When the eye makes too much fluid or the drainage system doesn't work properly, eye pressure may increase.

### **Open-angle glaucoma**

This is the most common form of glaucoma. The drainage angle formed by the iris and cornea remains open. But other parts of the drainage system don't drain properly. This may lead to a slow, gradual increase in eye pressure.

### **Acute angle-closure glaucoma**

This form of glaucoma happens when the iris bulges. The bulging iris partially or completely blocks the drainage angle. As a result, fluid can't circulate through the eye and pressure increases. Angle-closure glaucoma may happen suddenly or gradually.

### **Normal-tension glaucoma**

No one knows the exact reason why the optic nerve becomes damaged when eye pressure is healthy. The optic nerve may be sensitive or experience less blood flow. This limited blood flow may be caused by the buildup of fatty deposits in the arteries or other conditions that damage circulation. The buildup of fatty deposits in the arteries also is known as atherosclerosis.

### **Glaucoma in children**

A child may be born with glaucoma or develop it in the first few years of life. Blocked drainage, injury or an underlying medical condition may cause optic nerve damage.

### **Pigmentary glaucoma**

In pigmentary glaucoma, small pigment granules flake off from the iris and block or slow fluid drainage from the eye. Activities such as jogging sometimes stir up the pigment granules. That leads to a deposit of pigment granules on tissue located at the angle where the iris and cornea meet. The granule deposits cause an increase in pressure.

Glaucoma tends to run in families. In some people, scientists have identified genes related to high eye pressure and optic nerve damage.

**Risk factors**

Glaucoma can damage vision before you notice any symptoms. So be aware of these risk factors:

* High internal eye pressure, also known as intraocular pressure.
* Age over 55.
* Black, Asian or Hispanic heritage.
* Family history of glaucoma.
* Certain medical conditions, such as diabetes, migraine, high blood pressure and sickle cell anemia.
* Corneas that are thin in the center.
* Extreme nearsightedness or farsightedness.
* Eye injury or certain types of eye surgery.
* Taking corticosteroid medicines, especially eye drops, for a long time.

Some people have narrow drainage angles, putting them at increased risk of angle-closure glaucoma.

**Signs and symptoms**

The symptoms of glaucoma depend on the type and stage of the condition.

### **Open-angle glaucoma**

* No symptoms in early stages.
* Gradually, patchy blind spots in your side vision. Side vision also is called peripheral vision.
* In later stages, difficulty seeing things in your central vision.

### **Acute angle-closure glaucoma**

* Bad headache.
* Severe eye pain.
* Nausea or vomiting.
* Blurred vision.
* Halos or colored rings around lights.
* Eye redness.

### **Normal-tension glaucoma**

* No symptoms in early stages.
* Gradually, blurred vision.
* In later stages, loss of side vision.

### **Glaucoma in children**

* A dull or cloudy eye (infants).
* Increased blinking (infants).
* Tears without crying (infants).
* Blurred vision.
* Nearsightedness that gets worse.
* Headache.

### **Pigmentary glaucoma**

* Halos around lights.
* Blurred vision with exercise.
* Gradual loss of side vision.

**Diagnosis methods**

An eye care professional will review your medical history and do a comprehensive eye exam. Several tests may be done, including:

* Measuring intraocular pressure, also called tonometry.
* Testing for optic nerve damage with a dilated eye exam and imaging tests.
* Checking for areas of vision loss, also known as a visual field test.
* Measuring corneal thickness with an exam called pachymetry.
* Inspecting the drainage angle, also known as gonioscopy.

**Treatment options**

The damage caused by glaucoma can't be reversed. But treatment and regular checkups can help slow or prevent vision loss, especially if the disease is found in its early stages.

Treatment of glaucoma aims to lower intraocular pressure. Treatment options include prescription eye drops, oral medicines, laser treatment, surgery or a combination of approaches.

### **Eye drops**

Glaucoma treatment often starts with prescription eye drops. Some may decrease eye pressure by improving how fluid drains from the eye. Others decrease the amount of fluid the eye makes. Depending on how low the eye pressure needs to be, more than one eye drop may be prescribed.

Prescription eye drop medicines include:

* **Prostaglandins.** These increase the outflow of the fluid in the eye, helping to reduce eye pressure. Medicines in this category include latanoprost (Xalatan), travoprost (Travatan Z), tafluprost (Zioptan), bimatoprost (Lumigan) and latanoprostene bunod (Vyzulta).  
  Possible side effects include mild reddening and stinging of the eyes, darkening of the iris, darkening of the pigment of the eyelashes or eyelid skin, and blurred vision. This class of medicine is prescribed for once-a-day use.
* **Beta blockers.** These reduce the production of fluid in the eye, helping to lower eye pressure. Examples include timolol (Betimol, Istalol, Timoptic) and betaxolol (Betoptic S).  
  Possible side effects include difficulty breathing, slowed heart rate, lower blood pressure, impotence and fatigue. This class of medicine can be prescribed for once- or twice-daily use depending on your condition.
* **Alpha-adrenergic agonists.** These reduce the production of the fluid that flows throughout the inside of the eye. They also increase the outflow of fluid in the eye. Examples include apraclonidine (Iopidine) and brimonidine (Alphagan P, Qoliana).  
  Possible side effects include irregular heart rate; high blood pressure; fatigue; red, itchy or swollen eyes; and dry mouth. This class of medicine is usually prescribed for twice-daily use but sometimes can be prescribed for use three times a day.
* **Carbonic anhydrase inhibitors.** These medicines reduce the production of fluid in the eye. Examples include dorzolamide and brinzolamide (Azopt). Possible side effects include a metallic taste, frequent urination, and tingling in the fingers and toes. This class of medicine is usually prescribed for twice-daily use but sometimes can be prescribed for use three times a day.
* **Rho kinase inhibitor.** This medicine lowers eye pressure by suppressing the rho kinase enzymes responsible for fluid increase. It is available as netarsudil (Rhopressa) and is prescribed for once-a-day use. Possible side effects include eye redness and eye discomfort.
* **Miotic or cholinergic agents.** These increase the outflow of fluid from the eye. An example is pilocarpine (Isopto Carpine). Side effects include headache, eye pain, smaller pupils, possible blurred or dim vision, and nearsightedness. This class of medicine is usually prescribed to be used up to four times a day. Because of potential side effects and the need for frequent daily use, these medicines are not prescribed very often anymore.

Because some of the eye drop medicine is absorbed into the bloodstream, you may experience some side effects unrelated to your eyes. To minimize this absorption, close your eyes for 1 to 2 minutes after putting the drops in. You also may press lightly at the corner of your eyes near your nose to close the tear duct for 1 to 2 minutes. Wipe off any unused drops from your eyelid.

You may be prescribed multiple eye drops or need to use artificial tears. Make sure you wait at least five minutes in between using different drops.

### **Oral medicines**

Eye drops alone may not bring eye pressure down to the desired level. So an eye doctor also may prescribe oral medicine. This medicine is usually a carbonic anhydrase inhibitor. Possible side effects include frequent urination, tingling in the fingers and toes, depression, stomach upset, and kidney stones.

### **Surgery and other therapies**

Other treatment options include laser therapy and surgery. The following techniques may help to drain fluid within the eye and lower eye pressure:

* **Laser therapy.** Laser trabeculoplasty (truh-BEK-u-low-plas-tee) is an option if eye drops can't be tolerated. It also may be used if medicine hasn't slowed the progression of the disease. An eye doctor also may recommend laser surgery before using eye drops. It's done in the eye doctor's office. An eye doctor uses a small laser to improve the drainage of the tissue located at the angle where the iris and cornea meet. It may take a few weeks before the full effect of this procedure becomes apparent.
* **Filtering surgery.** This is a surgical procedure called a trabeculectomy (truh-bek-u-LEK-tuh-me). The eye doctor creates an opening in the white of the eye, which also is known as the sclera. The surgery creates another space for fluid to leave the eye.
* **Drainage tubes.** In this procedure, the eye surgeon inserts a small tube in the eye to drain excess fluid to lower eye pressure.
* **Minimally invasive glaucoma surgery (MIGS).** An eye doctor may suggest a MIGS procedure to lower eye pressure. This procedure generally require less immediate postoperative care and have less risk than trabeculectomy or using a drainage device. A MIGS procedure is often combined with cataract surgery. There are a number of MIGS techniques available.

After your procedure, you'll need to see your eye doctor for follow-up exams. And you may eventually need to undergo additional procedures if your eye pressure begins to rise or other changes happen in your eye.

### **Treating acute angle-closure glaucoma**

Acute angle-closure glaucoma is a medical emergency. If you're diagnosed with this condition, you'll need urgent treatment to reduce the pressure in your eye. This generally will require treatment with medicine and a laser or surgical procedures.

You may have a procedure called a laser peripheral iridotomy. The doctor creates a small hole in your iris using a laser. The hole allows fluid to flow through the iris. This helps to open the drainage angle of the eye and relieves eye pressure.

**Lifestyle and home remedies**

These tips may help control high eye pressure or promote eye health.

* **Eat a healthy diet.** Eating a healthy diet can help you maintain your health, but it won't prevent glaucoma from worsening. Several vitamins and nutrients are important to eye health, including zinc, copper, selenium, and antioxidant vitamins C, E and A.
* **Exercise safely.** Regular exercise may reduce eye pressure. Talk to your healthcare professional about an appropriate exercise program.
* **Limit your caffeine.** Drinking beverages with large amounts of caffeine may increase your eye pressure.
* **Sip fluids carefully.** Drink moderate amounts of fluids. Drinking a quart or more of any liquid within a short time may temporarily increase eye pressure.
* **Take prescribed medicine.** Using your eye drops or other medicines as prescribed can help you get the best possible result from your treatment. Be sure to use the eye drops exactly as prescribed. Otherwise, your optic nerve damage could get worse.

**Prevention tips**

These steps may help find and manage glaucoma in its early stages. That may help to prevent vision loss or slow its progress.

* **Get regular eye exams.** Regular eye exams can help find glaucoma in its early stages, before a lot of damage occurs. As a general rule, the American Academy of Ophthalmology recommends a comprehensive eye exam every 5 to 10 years if you're under 40 years old; every 2 to 4 years if you're 40 to 54 years old; every 1 to 3 years if you're 55 to 64 years old; and every 1 to 2 years if you're older than 65.  
  If you're at risk of glaucoma, you'll need screening more often. Ask a healthcare professional to recommend the right screening schedule for you.
* **Know your family's eye health history.** Glaucoma tends to run in families. If you're at increased risk, you may need screening more often.
* **Wear eye protection.** Serious eye injuries can lead to glaucoma. Wear eye protection when using power tools or playing sports.
* **Take prescribed eye drops regularly.** Glaucoma eye drops can greatly reduce the risk that high eye pressure will progress to glaucoma. Use eye drops as prescribed by a healthcare professional even if you have no symptoms.

**Alternative medicine**

Some alternative medicine approaches may help overall health, but none is an effective glaucoma remedy. Talk with an eye doctor about the possible benefits and risks.

* **Herbal remedies.** Some herbal supplements, such as bilberry extract, have been advertised as glaucoma remedies. But further study is needed to prove their effectiveness. Don't use herbal supplements in place of proven therapies.
* **Relaxation techniques.** Stress may trigger an attack of acute angle-closure glaucoma. Try to find healthy ways to cope with stress. Meditation and other techniques may help.
* **Marijuana.** Research shows that marijuana lowers eye pressure in people with glaucoma, but only for 3 to 4 hours. Other, standard treatments are more effective. The American Academy of Ophthalmology doesn't recommend marijuana for treating glaucoma.

**Prognosis**

Without treatment, glaucoma inevitably causes permanent vision loss and blindness. With treatment, it’s possible to slow the progress of the disease or stop it entirely. But because the range of possibilities can vary so widely, your eye specialist is the best person to talk to about this. They can tell you the likely outlook for your specific case and what you can do to help tilt the scales in your favor.

## **Living With**

### **What can I do to help myself if I have glaucoma?**

If you have glaucoma, the best thing you can do is follow your eye care specialist’s guidance on treating and managing this condition. They may also recommend making certain changes to your life, habits or routine. These can include:

* Not ignoring new symptoms or vision changes
* Reaching and maintaining a weight that’s healthy for you
* Staying physically active (but be sure to ask about which activities to avoid, as some can increase eye pressure)
* Seeing your provider as recommended

You should also see your provider if you notice new symptoms, if treatments aren’t as effective or if you have treatment side effects that are disrupting your life.

**When to see a doctor / red flag**

Angle-closure glaucoma usually develops quickly, so it needs immediate medical attention to prevent permanent damage and vision loss. The symptoms to watch for are the sudden onset or worsening of:

* Severe eye pain or pressure
* Headaches
* Double vision (diplopia) or blurred vision
* Nausea and vomiting that happen with eye pain/pressure
* Rainbow-colored halos around lights
* Vision loss of any kind
* Flashing lights in your vision
* Appearance or increase in visible floaters

## **Differential Diagnoses**

* Angle-Recession Glaucoma
* Anterior Ischemic Optic Neuropathy (AION)
* Aphakic and Pseudophakic Glaucoma
* Compressive Optic Neuropathy
* Drug-Induced Glaucoma
* Primary Angle-Closure Glaucoma
* Juvenile Glaucoma
* Lens-Particle Glaucoma
* Low-Tension Glaucoma
* Neovascular Glaucoma
* Phacolytic Glaucoma
* Phacomorphic Glaucoma
* Pigmentary Glaucoma
* Plateau Iris Glaucoma
* Posner-Schlossman Syndrome (PSS) (Glaucomatocyclitic Crisis)
* Primary Congenital Glaucoma
* Primary Open-Angle Glaucoma (POAG)
* Pseudoexfoliation Syndrome (Pseudoexfoliation Glaucoma)
* Toxic/Nutritional Optic Neuropathy
* Unilateral Glaucoma
* Uveitic Glaucoma

**Recent guidelines or updates**

#### Management Plan for Patients in Whom Therapy is Indicated

* The goal of treatment is to control the IOP in a target range and ensure the ONH/RNFL and visual fields are stable
* Target IOP is an estimate and must be individualized and/or adjusted during the course of the disease
* Set an initial target pressure of at least 25% lower than pretreatment IOP. Choosing a lower target IOP can be justified if there is more severe optic nerve damage, if the damage is progressing rapidly, or if other risk factors are present (e.g., family history, age, or disc hemorrhages).
* The IOP can be lowered by medical treatment, laser therapy, or incisional surgery (alone or in combination)
* Medical therapy is presently the most common initial intervention to lower IOP (see Table 4 of the POAG PPP for an overview of options available); consider balance between side effects and effectiveness in choosing a regimen of maximal effectiveness and tolerance to achieve the desired IOP reduction for each patient
* If progression occurs at the target pressure, undetected IOP fluctuations and adherence to the therapeutic regimen and recommendations for therapeutic alternatives should be discussed before adjusting target IOP downward
* Assess the patient who is being treated with glaucoma medication for local ocular and systemic side effects and toxicity
* Laser trabeculoplasty may be used as initial or adjunctive therapy in patients with POAG (see Table 5 of the POAG PPP). Laser trabeculoplasty is effective in lower IOP and may be performed to 180 degrees or 360 degrees of the angle.

#### Perioperative Care for Laser Trabeculoplasty Patients

* The ophthalmologist who performs surgery has the following responsibilities:
  + Obtain informed consent from the patient or patient's surrogate decision maker after discussing the risk, benefits, and expected outcomes of surgery
  + Ensure that the preoperative evaluation confirms that surgery is indicated
  + At least one IOP check immediately prior to surgery and within 30 minutes to 2 hours after surgery
  + Follow-up examination within 6 weeks of surgery or sooner if concern about IOP-related damage to the optic nerve

#### Perioperative Care for Incisional Glaucoma Surgery Patients

* The ophthalmologist who performs surgery has the following responsibilities:
  + Perform gonioscopy preoperatively, especially when considering the trabecular meshwork/Schlemm's canal-based MIGS (see Table 6 of the POAG PPP)
  + Obtain informed consent from the patient or patient's surrogate decision maker after discussing the risk, benefits, and expected outcome of surgery
  + Ensure that the preoperative evaluation accurately documents findings and indications for surgery
  + Prescribe topical corticosteroids in the postoperative period
  + Follow-up evaluation on the first postoperative day and at least once during the first 1 to 2 weeks to evaluate visual acuity, IOP, and status of the anterior segment
  + In the absence of complications, perform additional postoperative visits during a 3-month period to evaluate visual acuity, IOP, and status of the anterior segment
  + Schedule more frequent follow-up visits, as necessary, for patients with postoperative complications (flat or shallow anterior chamber, early bleb failure, increased inflammation, or Tenon's cyst)
  + Undertake additional treatments as necessary to improve aqueous flow into the bleb and lower IOP if evidence of bleb failure develops, including injection of antifibrotic agents, bleb massage, suture adjustment release or lysis, or bleb needling
  + Manage postoperative complications as they develop, such as repair of bleb leak or reformation of a flat anterior chamber
  + Explain that filtration surgery places the eye at risk for endophthalmitis for the duration of the patient's life and that if the patient has symptoms of pain and decreased vision and the signs of redness and discharge he or she should notify the ophthalmologist immediately

#### Patient Education For Patients with Medical Therapy

* Discuss diagnosis, severity of the disease, prognosis and management plan, and likelihood of lifelong therapy
* Educate about eyelid closure or nasolacrimal occlusion when applying topical medications to reduce systemic absorption
* Encourage patients to alert their ophthalmologist to physical or emotional changes that occur when taking glaucoma medications

## Primary Open-Angle Glaucoma (Follow-up Evaluation)

#### Exam History

* Interval ocular history
* Interval systemic medical history
* Side effects of ocular medications
* Review of pertinent medication use, including time of last administration

#### Physical Exam

* Visual acuity measurement
* Slit-lamp biomicroscopy
* IOP measurement
* Perform gonioscopy if there is a suspicion of angle-closure component, anterior chamber shallowing or anterior chamber angle abnormalities, or if there is an unexplained change in IOP. Perform gonioscopy periodically.
* ONH and visual field evaluation

#### Adjustment of Therapy

* Target IOP is not achieved and benefits of a change in therapy outweigh the risks
* Progressive optic nerve damage despite achieving the target IOP
* Patient's intolerant of the prescribed medical regimen
* Contraindications to individual medications develop
* Stable optic nerve status and low IOP occur for a prolonged period in a patient taking topical ocular hypotensive agents. Under these circumstances, a carefully monitored attempt to reduce the medical regimen may be appropriate.
* Downward adjustment of target pressure can be made in the face of progressive optic disc, imaging, or visual field change
* Upward adjustment of target pressure can be considered if the patient has been stable and if the patient either requires or desires less medication

#### Patient Education

* Educate about the disease process, the rationale and goals of intervention, the status of their condition, and relative benefits and risks of alternative interventions so that patients can participate meaningfully in developing an appropriate plan of action
* Patients considering keratorefractive surgery should be informed about the possible impact laser vision correction has on reducing contrast sensitivity and decreasing the accuracy of IOP measurements
* Patients with substantial visual impairment or blindness can be referred for and encouraged to use appropriate vision rehabilitation and social services

Among guideline recommendations for care:

* Examination of a person suspected of having POAG should include all aspects of a comprehensive eye and vision examination. Read the guideline to find out what to emphasize.
* Prostaglandin analogs should be considered as an initial therapy in patients with ocular hypertension or POAG, unless contraindicated.
* Patients prescribed topical intraocular pressure-lowering therapy may experience decreased tear film stability and elevated tear osmolarity.
* Pharmacological treatment of POAG should be used with caution during pregnancy and lactation.
* Selective laser trabeculoplasty should be considered as an initial/alternative or additive therapy to medication for intraocular pressure (IOP) control.
* The frequency and scope of follow-up examinations of patients should be individualized, based on the severity and stability of their disease and should occur at regular intervals to monitor progression and treatment efficacy.
* Early medical treatment should be considered for individuals with ocular hypertension who are at moderate or high risk of developing POAG.
* Eye doctors should be persistent in providing education and training to patients to improve adherence/compliance with therapy.
* Patients prescribed pharmacological treatment should receive instructions on eyedrop instillation.
* Ocular telehealth can provide increased access to care but should not be used alone or for the assessment or management of moderate or advanced disease.

**Epidemiology data**

* Glaucoma is the most common cause of irreversible blindness.
* Based on prevalence studies, it is estimated that 79.6 million individuals will have glaucoma in 2020. This number is likely to increase to 111.8 million individuals in 2040.
* At least, half of those with glaucoma are unaware that they are affected. In some developing countries, 90% of glaucoma is undetected.
* In many cases, glaucoma may be asymptomatic.
* It is estimated that more than 11 million individuals will be bilaterally blind due to glaucoma in 2020 (around 13% of the cases).
* In most cases, blindness can be prevented with appropriate control and treatment.
* In the USA, blindness is the third most feared health problem, after cancer and cardiac attacks.
* Unfortunately, many individuals are unaware of the existence of glaucoma.
* A better awareness could prevent visual disability in many people.

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**Hemianopia**

**Definition and description**

There are different types of hemianopia, including homonymous hemianopia and heteronymous hemianopia. Each type is classified by what part of the visual field is affected.

Because hemianopia stems from an event or complication, hemianopia is considered a brain condition, not an eye condition.

## Types of Hemianopia

The visual pathway is part of the central nervous system (CNS). It is made up of several vital neurons that turn light into visual stimuli, including:

* Retina (thin tissue layer in the back of the eye that translates light to the optic nerve)
* Optic nerves (communication pathway between the eye and the brain)
* Optic chiasm (joining together of the optic nerves of both eyes)
* Visual cortex (primary area of the brain that receives and processes light from the retina)

During visual processing, the optic nerve from the right eye joins with the optic nerve from the left to form an X-shaped structure called the optic chiasm. The optic nerves then cross paths and continue on the optic tract to the brain.

The brain is divided into two halves, which are the right side and the left side:

* The left side is responsible for vision in the right visual field of each eye
* The right side is responsible for vision in the left visual field of each eye

The type of hemianopia depends on the location of the disturbance in the brain or visual pathway.

### **Homonymous Hemianopia**

Homonymous hemianopia (also known as homonymous hemianopsia or HH) is a symptom that makes you see only one side ― right or left ― of the visual field of each of your eyes. “Homonymous” in this context means “the same side of both eyes,” and “hemianopia” (sometimes spelled “hemianopsia”) means “half vision loss.”

Your visual field is everything you see when your eyes are open. When your vision works correctly, everything on the right side of your visual field goes to the left side of your brain, and everything on the left side goes to the right side of your brain. Your visual system has a special setup to make that happen.

Each optic nerve carries visual information from both sides until they reach a point in your brain called the optic chiasm (pronounced “KY-azm”). Once the nerve fibers get there, some of the fibers change directions. Nerve fibers from both eyes that handle the left side of your vision go to the right side of your brain and vice versa.

HH affects signals from the same side of both eyes traveling together after passing the chiasm. That’s why HH causes vision loss on the same side of both eyes. Experts classify it as a visual field defect, and the conditions that cause it all affect your brain. It’s usually sudden, but some conditions can make it develop more gradually.

## **Possible Causes**

### **What are the most common causes of homonymous hemianopia?**

HH means something is disrupting your visual system on one side of your brain. Between 42% and 89% of HH cases happen because of these three life-threatening emergency conditions:

* Strokes.
* Transient ischemic attacks (TIAs).
* Brain bleeds (intracranial hemorrhages).

#### **Strokes and TIAs**

Strokes happen when something disrupts blood flow to part of your brain. Without blood flow, the affected brain areas stop working correctly and start dying. When this is severe or lasts too long, it causes permanent brain damage. These can happen because of a blockage (ischemic stroke) or bleeding (hemorrhagic stroke).

People often call TIAs “mini-strokes,” but they aren’t necessarily smaller. They’re like an ischemic stroke, but the symptoms are temporary (they usually stop if you sit down or rest). But having a TIA greatly increases your risk of having a stroke within the next few days. That’s why a TIA is also a medical emergency, just like a stroke.

#### **Brain bleeds**

Bleeding inside your brain is dangerous because the blood has nowhere to go. Over time, the buildup of blood can press on and damage your brain.

#### **Other conditions that can cause homonymous hemianopia**

Many other brain conditions can also cause HH when they damage vision-related areas. Some of these conditions are dangerous or life-threatening. Others are relatively minor, and the risk of permanent damage is minimal.

Examples of other conditions that can cause HH include:

* Brain infections and inflammation (such as encephalitis or neurosyphilis).
* Brain lesions.
* Brain tumors (including cancer).
* Concussions and traumatic brain injuries (TBIs).
* Degenerative brain diseases like Alzheimer’s disease or Creutzfeldt-Jakob disease (CJD).
* Inflammatory and autoimmune conditions like multiple sclerosis (MS) or neuromyelitis optica (NMO).
* Metabolic conditions or effects, like low blood sugar (hypoglycemia).
* Migraines (especially migraines with auras).
* Seizures and epilepsy.

## **Care and Treatment**

### **How is homonymous hemianopia treated?**

The treatments for HH vary widely depending on the underlying cause. Some examples of treatment options for specific conditions that can cause homonymous hemianopia include:

* Ischemic stroke or TIA: The main treatments for ischemic stroke restore blood flow with medications, like anticoagulants or clot-busting (thrombolytic) drugs, or specific medical procedures like thrombectomy.
* Brain bleeds and hemorrhagic stroke: Treatments usually involve surgery (like craniectomy or craniotomy) to relieve pressure inside your skull and repair the affected blood vessel, if possible.
* Migraines: Treatments usually include preventive medications that keep migraines from happening or abortive (“rescue”) medications to stop them when they happen. Dozens of medications fall into one or both of those categories.
* Seizures and epilepsy: Antiseizure medications can stop seizures, prevent them or both. Epilepsy surgery is also an option if medications aren’t effective or helpful.

Something to remember is that treatments that work for one cause may not work for others, or they could do more damage. Because there are so many treatment options and other factors in play, your healthcare provider is the best person to tell you about the treatment options for your specific case.

### **Can homonymous hemianopia be prevented?**

HH is impossible to prevent, and it happens unpredictably. But you can reduce your risk of having some of the conditions that cause it if you take the following steps:

* See a healthcare provider for an annual physical or checkup to improve the odds of early detection of conditions that could cause HH.
* Wear safety gear like helmets and seatbelts to reduce the risk of head injuries.
* Reach and maintain a weight that’s healthy for you to prevent or delay a stroke or similar conditions.
* Manage chronic conditions to prevent issues or events that could cause HH, like taking epilepsy medicine to prevent seizures or anticoagulants to prevent clots that could cause a stroke.

## **When To Call the Doctor**

### **When should homonymous hemianopia be treated by a doctor or healthcare provider?**

If you’ve never experienced homonymous hemianopsia before, you should react like it’s a medical emergency. HH is most likely a symptom of a life-threatening emergency condition like a stroke or brain bleed.

The best and safest course of action is to call 911 (or your local emergency services number) immediately. You shouldn’t try to drive yourself to get medical care because the loss or disruption in your vision can make driving dangerous to yourself and others.

If you’ve experienced HH before and have a diagnosed condition that can cause it, how you respond can vary slightly. You should ask your healthcare provider what to do if you have it again. They can give you more specific guidance on when this symptom needs immediate medical attention.

But when in doubt, the safest thing to do is to get medical attention. Many conditions that cause HH are time-sensitive, and seconds can make a huge difference.

### **Heteronymous Hemianopia**

Heteronymous hemianopia is a bilateral visual field defect on the opposite sides of each eye.

This type of hemianopia results from a lesion on the optic chiasm, the X shape structure where the optic nerves meet and cross.

The types of heteronymous hemianopia include:

* Bitemporal hemianopia is vision loss in the outer part of the visual field in each eye
* Binasal hemianopia is vision loss in the inner part of the visual field in each eye

## **Common Symptoms**

Depending on the underlying cause of hemianopia, it can be either temporary or permanent.

Symptoms of homonymous hemianopia range from mild to severe and include:

* Disturbed site and missing objects in the visual field
* Blurry or double vision
* Dimmed vision
* Trouble with night vision
* Visual hallucinations
* Visual neglect is when you fail to notice on one side of your visual field

Heteronymous hemianopia symptoms are less severe and usually only cause depth perception and peripheral vision problems.

**What Causes Hemianopia?**

Stroke is the most common cause of homonymous hemianopia and accounts for 69.7% of cases.1

Other causes of hemianopia include:

Brain injuries

Bleeding in the brain

* Brain tumors
* Infection in the brain
* Multiple sclerosis
* Alzheimer's disease
* Epilepsy
* Invasive surgical procedures

## **Symptoms of hemianopia**

Losing part of your visual field isn’t as obvious as you might think. However, there are some symptoms that may reveal hemianopia, including:

* Double vision
* Inability to see objects located in the affected area of the visual field, such as cars in other lanes while driving, or food on a specific area of a plate
* Visual hallucinations that appear as lights and various shapes
* Turning the head away from the side affected by hemianopia
* Bumping into things on the affected side
* Disorientation in crowded environments
* Drifting away from the affected visual field when walking
* Reading only part of a block of text because the other part is “missing”

**Diagnosis methods**

Though hemianopia is a problem with the brain and not the eyes, you will need an eye exam to diagnose the condition. Your eye doctor will have you perform a visual field test that notes the location and size of the affected areas of your visual field.

The exam also includes having the doctor examine the inside of your eyes. This allows them to check the health of the retinas , macula and optic discs.

Additionally, the doctor will check the internal pressure of your eyes to look for other possible causes of peripheral (side) vision loss , such as glaucoma .

Besides an eye exam, your doctor will want you to obtain a head MRI (magnetic resonance imaging) to assess the health of your brain.

## **How is Hemianopia Treated?**

Hemianopia treatment is dependent on the underlying cause of the visual disturbance. The condition may correct itself over time with spontaneous recovery from a brain injury, stroke, or tumor removal.

Some cases of hemianopia may be permanent and need ongoing treatment to expand your visual world.

These treatment options include:

* Vision restoration therapy to help with reading and dealing with your environment
* Visual rehabilitation to maximize the use of remaining vision and eye movement patterns
* Visual assist devices, including prism glasses
* Using a driving simulator to determine if you are safe to drive
* Learning how to make quick eye and head movements toward the affected side
* Use a straight edge to help direct your eyes when reading

### **Can You Recover from Hemianopia?**

Visual recovery from hemianopia depends on the underlying cause of the blind side and your ability to heal from injury or illness.

Permanent loss of half of the visual fields can affect the quality of life, including:

* Not being able to drive
* Difficulty reading
* Trouble moving around in your environment
* Social isolation
* Anxiety and depression

**prognosis**

Visual loss in the setting of stroke suggests a poor prognosis, with the rate of improvement ranging widely from 17% to 67%. Certain studies indicate that only 18% of patients with homonymous hemianopsia regain their vision within 28 to 30 days of the inciting event. In contrast, others report spontaneous improvement in 46% to 67% of cases within 1 month of an ischemic stroke. The chances of visual recovery diminish over time, with the likelihood of regaining visual function notably low after 6 months.

Patients with bitemporal hemianopsia attributed to pituitary tumors typically experience visual field improvement in 79% to 95% of cases following resection. However, the extent of recovery may hinge on factors such as retinal nerve fiber layer thickness, preoperative deficit severity, duration of visual symptoms, tumor size, extent of resection, and patient age.

**Possible complications**

Complications associated with hemianopsia, including impaired balance, heightened risk of falls and injury, loss of independence, reading difficulties, impaired visual scanning, increased risk of depression, and reduced employment opportunities, can significantly impact the quality of life in patients.

**Differential diagnosis**

The differential diagnosis of hemianopsia relates closely to the location and the underlying etiology. The following list contains the differential diagnoses for hemianopsia:

* Cerebral infarction
* Intracranial hemorrhage
* Saccular aneurysm
* Mass lesions, including primary and metastatic tumors
* Pituitary adenoma
* Craniopharyngioma
* Meningioma
* Central nervous system lymphoma or other primary malignancy
* Altitudinal hemianopia
* Trauma
* Inflammatory conditions, including demyelinating disorders
* Severe hyperglycemia
* Degenerative neurological conditions
* Migraine
* Transient ischemia attack
* Seizure or status epilepticus
* Immunoglobulin G4–related disease

**Epidemiology**

The occurrence of hemianopsia varies depending on the underlying pathological condition. In adults, stroke stands out as the predominant cause, accounting for 69.7% of cases, with an average patient age of 58.Conversely, in children, neoplasms contribute to 39% of cases, cerebrovascular disease to 25%, and trauma to 19%. An extensive review involving 904 patients with homonymous hemianopsia highlights that stroke primarily affects the occipital lobe in 54% of patients, followed by involvement of the optic radiations in 33% and optic tracts in 6% of patients.

In non-stroke-related homonymous hemianopsia, damage can occur at various locations along the visual pathway. Specifically, 24% affect the occipital lobe, 31% impact the optic radiations, 19% involve the optic tract, and 25% affect multiple locations. Pituitary adenomas are the most frequently encountered cause of bitemporal hemianopsia. A systematic review based on radiographic and autopsy studies estimates the overall prevalence of pituitary tumors to be 16.7%. Conversely, occlusive cerebrovascular disease or occipital infarction is identified as the most common cause of homonymous hemianopia with macular sparing

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**HORNER SYNDROME**

ALTERNATIVE NAMES: Horner’s syndrome is also known as “oculosympathetic paresis”, “Bernard-Horner syndromes (BH)”, and “oculosympathetic palsy”.

**DEFINITION / DESCRIPTION**

Horner’s syndrome occurs as a result of a lesion along the oculo-sympathetic pathway. Clinical features include ipsilateral miosis, ptosis, anhidrosis, enophthalmos, and loss of ciliospinal reflex. Inverse ptosis (upside-down ptosis), a condition where the lower eyelid elevates due to underlying denervated tarsal muscle, may also be seen. It is worth noting that the enophthalmos in Horner syndrome is not a true enophthalmos. It results due to evident narrowing of vertical palpebral fissure height, producing an apparent sunken appearance of the globe.

Prior knowledge of the sympathetic pathway is important to localize the lesion and understand the rationale behind the clinical presentation of Horner syndrome.

The sympathetic pathway comprises a three–order neuronal chain. The first–order neuron arises from the posterolateral hypothalamus and synapses at the C8-T2 level in the intermediolateral column of the spinal cord, also called the ciliospinal center of Budge. The second-order neurons now travel across the apex of the lung and along the sympathetic chain, ultimately synapsing at the level of the superior cervical ganglion. The third-order neurons (postganglionic fibers) travel with the internal carotid artery (ICA) through the cavernous sinus and then along the abducens nerve. It then follows the first division of the trigeminal nerve as the nasociliary nerve and later as the long ciliary nerves to supply the dilator pupillae.

Anhidrosis is typically present in cases of central (1 order) or preganglionic (2 order) lesions. Although the classical clinical features are sufficient to diagnose Horner syndrome, several pharmacological tests are available to confirm a case of suspected Horner syndrome.

*Drug* *Mechanism of Action*

Anisocoria due to Horner syndrome associated with pain should be treated as an emergency warranting immediate recognition. The most important cause of a painful Horner’s syndrome is ICA dissection.

Children with anisocoria must be evaluated for birth trauma. It is the most common cause of congenital Horner’s syndrome. If no evidence of trauma is found, a dedicated work-up to rule out neuroblastoma is mandated in children. Appropriate imaging, including Magnetic Resonance Imaging (MRI) head, neck, chest, and abdomen, should be carried out for timely detection and appropriate management.

**Cholinergic Medications**

Cholinergic medications can cause miosis of the pupil through activation of the sphincter pupillae. The most common eye drop is pilocarpine, which has historically been used to treat glaucoma.

**Posterior Synechiae**

If posterior synechiae (abnormal connections between the iris and the lens) develop, the pupil may not be able to dilate fully. This can be caused by uveitis, angle-closure glaucoma, or other inflammatory conditions. The best way to evaluate for posterior synechiae is with a slit lamp examination. In this case, the underlying condition needs to be treated.

**CAUSES**

**Causes of Anisocoria Greater in Bright Light**

**Third Nerve Palsy**

The parasympathetic fibers run along the periphery of the third nerve. Thus, compressive lesions of the third nerve would affect the parasympathetic fibers. This inhibits the signal to the sphincter pupillae, causing unopposed dilation of the pupil on the affected side. A third nerve palsy can be differentiated from other causes if the oculomotor component is affected. There would be significant ptosis of the affected upper eyelid due to inhibition to the levator palpebrae superioris. The eye would also be abducted and depressed, also known as "down and out." This is due to the unopposed action of the lateral rectus and superior oblique, which are the two extraocular muscles not innervated by the third cranial nerve. A very concerning compressive lesion would be an aneurysm, which can be acutely fatal.

**Migraine Headache**

A migraine headache can cause anisocoria associated with pain. The pupil will typically be dilated, and the dilation often resolves after the headache has subsided.

**Cycloplegic Medications**

Cycloplegic medications cause paralysis of the sphincter pupillae, which in turn causes unopposed dilation. This pupil will, at most, minimally constrict with light and, much of the time, will not react at all due to paralysis. Pilocarpine will not be effective in causing this pupil to constrict. Cycloplegic eye drops include cyclopentolate, atropine, and tropicamide. The same effect can be achieved if a patient touches an anticholinergic medication and then touches the eye. Getting a history of cycloplegic medication use is the best way to make this diagnosis.

**Sympathomimetics**

Eye drops that cause activation of the dilator pupillae can cause mydriasis of the pupil. There still should be some reactivity of the pupil. The dilation is not as great as with cycloplegic medications.

**Adie Tonic Pupil**

In this condition, the larger pupil fails to constrict to the extent of the other pupil. However, when the eye accommodates, the pupil has greater constriction. This cause of anisocoria can be diagnosed with dilute pilocarpine, which causes significant constriction of the larger pupil. Other causes of a dilated pupil typically do not respond to dilute pilocarpine, helping confirm the diagnosis of Adie tonic pupil.

**Traumatic Mydriasis**

Damage to the pupillary sphincter due to trauma or intraocular surgery may leave the pupil dilated and potentially with an irregular shape. The pupil may then be unable to constrict. The best way to diagnose traumatic mydriasis is to get a complete history of ocular trauma and eye surgeries and then perform a slit lamp exam. Mydriasis due to trauma may improve over time or remain permanent. An irregular pupil after intraocular surgery has a higher likelihood of remaining permanent.

**Signs and symptoms**

Horner syndrome usually affects only one side of the face. Common signs and symptoms include:

* A persistently small pupil (miosis)
* A notable difference in pupil size between the two eyes (anisocoria)
* Little or delayed opening (dilation) of the affected pupil in dim light
* Drooping of the upper eyelid (ptosis)
* Slight elevation of the lower lid, sometimes called upside-down ptosis
* Sunken appearance of the affected eye
* Little or no sweating (anhidrosis) on the affected side of the face

Signs and symptoms, particularly ptosis and anhidrosis, may be subtle and difficult to detect.

### **Children**

Additional signs and symptoms in children with Horner syndrome may include:

* Lighter iris color in the affected eye of a child under the age of 1
* Change in color on the affected side of the face that would typically appear from heat, physical exertion or emotional reactions

**Diagnosis methods**

In addition to a general medical examination, your doctor will likely conduct tests to determine the nature of your symptoms and identify a possible cause.

### **Tests to confirm Horner syndrome**

Your doctor may be able to diagnose Horner syndrome based on your history and an assessment of your symptoms.

An eye specialist (ophthalmologist) may also confirm a diagnosis by putting a medicated eye drop in both eyes — either a drop that will dilate the pupil of a healthy eye or a drop that will constrict the pupil in a healthy eye. By comparing the reactions in the healthy eye with that of the suspect eye, the doctor can determine whether nerve damage is the cause of problems in the suspect eye.

### **Tests to identify the site of nerve damage**

The nature of your symptoms may help your doctor narrow the search for the cause of Horner syndrome. Your doctor may also conduct additional tests or order imaging tests to locate the lesion or irregularity disrupting the nerve pathway.

Your doctor may administer a type of eye drop that will significantly dilate the healthy eye and little dilation of the affected eye if Horner syndrome is caused by a third-order neuron irregularity — a disruption somewhere in the neck or above.

Your doctor may order one or more of the following imaging tests to locate the site of a probable irregularity causing Horner syndrome:

* Magnetic resonance imaging (MRI), a technology that uses radio waves and a magnetic field to produce detailed images
* Magnetic resonance angiography (MRA), which is used to evaluate blood vessels
* Chest X-ray
* Computerized tomography (CT), a specialized X-ray technology

**Treatment options**

There's no specific treatment for Horner syndrome. Often, Horner syndrome disappears when an underlying medical condition is effectively treated.

**Prevention tips**

As so many underlying conditions can potentially cause Horner syndrome, there’s no way to prevent it from developing.

However, in cases of Horner syndrome related to trauma (such as carotid artery dissection), taking safety measures to avoid injuring your neck and following dissection precautions can help you avoid developing the syndrome.

**Prognosis**

The prognosis (outlook) of Horner syndrome depends on the underlying cause. The characteristic symptoms of Horner syndrome generally don’t have a significant impact on your quality of life or vision.

If the underlying cause is a chronic (long-lasting) condition, such as multiple sclerosis, Horner syndrome may also be chronic. If it results from something temporary and treatable, such as an ear infection, it’ll likely go away once the infection is treated.

**POSSIBLE COMPLICATIONS**

Anisocoria itself is unlikely to cause significant complications, although some do exist. A larger pupil may cause light sensitivity and visual aberrations. A smaller pupil may cause worsened visualization through a cataract, difficulty viewing the fundus during the posterior exam, or difficulty in cataract surgery. The main complication of anisocoria is not the difference in pupil size but the complications of the underlying condition itself.

**When to see a doctor / red flag**

If you have symptoms of Horner syndrome, such as one drooping upper eyelid, mismatched pupil sizes and lack of sweating on the same side of your face, see a healthcare provider as soon as possible.

**Differential diagnosis**

## Central (First-Order Neuron) Lesions

Lesions affect the hypothalamus, brainstem, or cervical spinal cord (C8–T2). Common causes include:

* Stroke: Lateral medullary syndrome (Wallenberg syndrome)
* Brainstem tumors: Glioma, vascular malformations
* Demyelinating diseases: Multiple sclerosis
* Syringomyelia or syringobulbia (cystic cavities in spinal cord or brainstem)
* Trauma: Cervical spinal cord injury
* Arnold-Chiari malformation
* Basal meningitis (e.g., syphilis)
* Pituitary tumors

2. Preganglionic (Second-Order Neuron) Lesions

Lesions in the thoracic spinal cord, thoracic outlet, or lower neck affecting sympathetic fibers before they synapse in the superior cervical ganglion. Causes include:

* Pancoast tumor: Apical lung carcinoma compressing the sympathetic chain
* Thoracic outlet syndrome: Cervical rib, subclavian artery aneurysm
* Mediastinal tumors or lymphadenopathy (e.g., lymphoma, tuberculosis)
* Birth trauma or brachial plexus injury
* Surgical trauma: Radical neck dissection, chest tube placement, jugular vein cannulation
* Aortic aneurysm or dissection
* Thyroid malignancies

3. Postganglionic (Third-Order Neuron) Lesions

Lesions distal to the superior cervical ganglion affecting the sympathetic fibers traveling along the internal carotid artery and into the orbit. Causes include:

* Internal carotid artery dissection or thrombosis (often painful, with ipsilateral headache or neck pain)
* Carotid artery aneurysm or invasive tumors at skull base
* Cavernous sinus lesions: Thrombosis, aneurysm, tumors
* Cluster headaches (transient Horner syndrome during attacks)
* Raeder paratrigeminal syndrome: Horner syndrome with trigeminal nerve pain and sensory loss
* Trauma or surgery near the skull base or neck
* Invasive pituitary tumors or parasellar tumors

4. Other Causes in Children

* Birth trauma (most common cause in pediatrics)
* Neuroblastoma (a common pediatric malignancy causing Horner syndrome)
* Neck trauma or surgical injury

5. Other Conditions to Consider (Mimics or Related Syndromes)

* Physiologic anisocoria
* Adie pupil
* Argyll Robertson pupil
* Third nerve palsy
* Iris sphincter damage
* Drug-induced miosis or ptosis (e.g., topical miotics, systemic medications)

**Epidemiology data**

Horner syndrome is uncommon, occurring with a frequency of approximately 1 per 6,000. It may occur at any age or with any ethnic group.

* The frequency of Horner syndrome is estimated at approximately 1 in 6,250 to 1 in 6,000 in the general population .
* The age- and sex-adjusted incidence in a population-based study was 4.2 per 100,000 per year, with incidence increasing with age .

Pediatric Epidemiology:

* The birth prevalence of congenital Horner syndrome is estimated to be 1 in 6,250 newborns .
* The incidence of Horner syndrome in patients younger than 19 years is estimated at 1.42 per 100,000.
* A nationwide study in South Korea reported a cumulative incidence of 2.12 per 100,000 in the pediatric population .
* The peak incidence in children occurs at 0–4 years of age.
* In children, trauma (including birth trauma) is the most common cause, and acquired cases often require thorough investigation due to potentially serious underlying causes like neuroblastoma.

Adult Epidemiology:

* In adults, the cumulative incidence was 2.95 per 100,000 in the South Korean study .
* The peak incidence in adults occurs at 50–54 years of age .
* In adult cases, the most common etiologies include idiopathic (26%), internal carotid artery (ICA) dissection (19%), and stroke (15%) . Thyroid tumors are also a common tumor-related cause in adults.

Age and Sex Distribution:

* Horner syndrome can occur at any age and in any ethnicity .
* While there are no significant differences based on sex in overall incidence, a South Korean study found that 59.7% of pediatric patients and 51.0% of adult patients newly diagnosed with Horner syndrome were male

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**Keratoconus**

**Definition and description**

Keratoconus is a condition in which there is a thinning and cone-shaped deformation of the cornea, distorting vision and usually developing in younger individuals. It can be managed with contact lenses or surgery.

Keratoconus (ker-uh-toe-KOH-nus) is an eye condition in which the clear, dome-shaped front of the eye, called the cornea, gets thinner, steeper and bulges outward into a cone shape.

A cone-shaped cornea causes blurred vision and may cause sensitivity to light and glare. Keratoconus usually affects both eyes. However, it can affect one eye more than the other. It generally begins to affect people between the late teens and 30 years of age. The condition may progress slowly for 10 years or longer.

In the early stages of keratoconus, you might be able to correct vision problems with glasses or soft contact lenses. Later, you may have to be fitted with rigid, gas permeable contact lenses or other types of lenses, such as scleral lenses. If your condition gets worse, you may need a cornea transplant.

A procedure called corneal collagen cross-linking may help to slow or stop keratoconus from progressing, possibly preventing the need for a future cornea transplant. This treatment may be offered in addition to the vision correction options above.

**Causes**

No one knows what causes keratoconus, although genetic and environmental factors are thought to be involved. Around 1 in 10 people with keratoconus also have a parent with the condition.

**Risk factors**

These factors can increase the risk of developing keratoconus:

* Having a family history of keratoconus.
* Rubbing the eyes vigorously.
* Having certain conditions, such as retinitis pigmentosa, Down syndrome, Ehlers-Danlos syndrome, Marfan syndrome, hay fever and asthma.

**Signs and symptoms**

Symptoms of keratoconus may change as the disease progresses. They include:

* Blurred or distorted vision.
* Increased sensitivity to bright light and glare, which can cause problems with night driving.
* A need for frequent changes in eyeglass prescriptions.
* Sudden worsening or clouding of vision.

**Diagnosis methods**

To diagnose keratoconus, an eye doctor will review a medical and family history and do an eye exam. Other tests also may be done to find out more about the shape of the cornea.

Tests to diagnose keratoconus include:

* **Eye refraction.** This test uses special equipment that measures the eyes. It may involve looking through a device that contains wheels of different lenses, called a phoropter. This device helps judge which combination offers the sharpest vision. Some eye doctors may use a hand-held instrument called a retinoscope to check the eyes.
* **Slit-lamp examination.** This test involves directing a vertical beam of light on the surface of the eye and using a low-powered microscope to view the eye. The eye doctor evaluates the shape of the cornea and looks for other potential problems in the eye.
* **Keratometry.** This exam involves focusing a circle of light on the cornea and measures the reflection. This determines the basic shape of the cornea.
* **Computerized corneal mapping.** Special photographic tests, such as corneal tomography and corneal topography, record images to create a detailed shape map of the cornea. Corneal tomography also can measure the thickness of the cornea. This type of testing can often detect early signs of keratoconus before the disease is visible by slit-lamp examination.

**Treatment options**

Treatment for keratoconus depends on the severity of the condition and how quickly the condition is progressing. Generally, there are two approaches to treating keratoconus: slowing the progression of the disease and improving vision.

If keratoconus is progressing, corneal collagen cross-linking may be recommended to slow it or stop it from getting worse. This treatment aims to stabilize the structure of the cornea. It may decrease the bulging of the cornea and help achieve better vision with glasses or contact lenses. This treatment also has the potential to prevent needing a cornea transplant in the future.

Improving vision depends on the severity of keratoconus. Mild to moderate keratoconus can be treated with eyeglasses or contact lenses. This will likely be a long-term treatment, especially if the cornea becomes stable with time or from cross-linking.

In some people with keratoconus, the cornea becomes scarred with advanced disease. For others, wearing contact lenses becomes difficult. In these people, cornea transplant surgery might be necessary.

### **Lenses**

* **Eyeglasses or soft contact lenses.** Glasses or soft contact lenses can correct blurry or distorted vision in early keratoconus. But people often need to change their prescription for eyeglasses or contacts as the shape of their corneas change.
* **Hard contact lenses.** Hard contact lenses are often the next step in treating more advanced keratoconus. Hard lenses include rigid, gas permeable types. Hard lenses may feel uncomfortable at first, but many people adjust to wearing them and they can provide excellent vision. This type of lens can be made to fit the corneas.
* **Piggyback lenses.** If rigid lenses are uncomfortable, an eye doctor may recommend "piggybacking" a hard contact lens on top of a soft one.
* **Hybrid lenses.** These contact lenses have a rigid center with a softer ring around the outside for increased comfort. People who can't tolerate hard contact lenses may prefer hybrid lenses.
* **Scleral lenses.** These lenses are useful for very irregular shape changes in the cornea in advanced keratoconus. Instead of resting on the cornea like traditional contact lenses do, scleral lenses sit on the white part of the eye, called the sclera, and vault over the cornea without touching it.

If you're using rigid or scleral contact lenses, make sure to have them fitted by an eye doctor with experience in treating keratoconus. You'll also need to have regular checkups to determine whether the lenses still fit well. An ill-fitting lens can damage the cornea.

### **Therapies**

* **Corneal cross-linking.** In this procedure, the cornea is saturated with riboflavin eye drops and treated with ultraviolet light. This causes cross-linking of the cornea, which stiffens the cornea to prevent further shape changes. Corneal cross-linking may help to reduce the risk of progressive vision loss by stabilizing the cornea early in the disease.

### **Surgery**

Surgery may be necessary if there is corneal scarring, extreme thinning of the cornea, poor vision with the strongest prescription lenses or an inability to wear any type of contact lenses.

Depending on the location of the bulging cone and the severity of the condition, surgical options include:

* **Intrastromal corneal ring segments (ICRS).** For mild to moderate keratoconus, an eye doctor may recommend inserting small synthetic rings in the cornea. This treatment can help flatten the cornea, which can help improve vision and make contact lenses fit better. Sometimes this procedure is done in combination with corneal cross-linking.
* **Cornea transplant.** If there is corneal scarring or extreme thinning, a cornea transplant may be needed. Depending on the situation, an eye doctor may recommend replacing all or part of the cornea with healthy donor tissue. A cornea transplant is known as a keratoplasty.

Cornea transplant for keratoconus generally is very successful. Possible complications include graft rejection, poor vision, infection and astigmatism. Astigmatism is often managed by wearing hard contact lenses again, which is usually more comfortable after a cornea transplant.

**Prevention tips**

No, you can’t prevent keratoconus. If you have a condition associated with keratoconus, you may be able to reduce your risk by doing your best to avoid rubbing your eyes.

**Prognosis**

With treatment, the outlook for someone with keratoconus is good.

If your vision or prescription is different in each eye, you might find yourself dealing with balance issues. Speak to your provider about this. They’ll help you find a solution.

Every person is different. Some people have mild cases of keratoconus that don’t progress. Other people have cases that do progress. No one can predict what will happen in every case.

### **Can keratoconus damage vision?**

Untreated keratoconus can lead to permanent vision loss. The changes to the cornea make it difficult for your eye to focus with or without eyeglasses or standard soft contact lenses.

In addition, keratoconus can be dangerous if you have laser vision correction surgery such as LASIK because the surgery can make keratoconus worse. If you’re thinking about this kind of surgery, your eye surgeon will do an exam to see if you’re a candidate. If you have even a small degree of keratoconus, you shouldn’t have LASIK, unless your provider specifically recommends it.

**Possible complications**

In some situations, the cornea may swell quickly and cause sudden reduced vision and scarring of the cornea. This is caused by a condition in which the inside lining of the cornea, called Descemet's membrane, breaks down. This causes fluid to enter the cornea, a condition known as hydrops. The swelling usually goes down by itself, but a scar may form that affects vision.

Advanced keratoconus also may cause the cornea to become scarred, particularly where the cone is most prominent. A scarred cornea causes worsening vision problems and may require cornea transplant surgery.

**When to see a doctor / red flag**

You should always contact your eye care provider when you have changes in vision. If you have keratoconus, you’ll probably need to see your provider on a regular basis. Keep your appointments.

**Differential diagnosis**

Pellucid Marginal Degeneration (PMD): A bilateral, non-inflammatory ectatic disorder characterized by thinning in the inferior peripheral cornea, often sparing the central cornea, unlike the central or paracentral thinning seen in keratoconus. PMD typically presents with against-the-rule astigmatism and a "crab-claw" pattern on corneal topography.

* Keratoglobus: A rare, bilateral condition involving diffuse corneal thinning and globular protrusion of the entire cornea, rather than the localized cone-shaped protrusion in keratoconus. It usually presents with more extensive thinning and may be congenital or acquired.
* Contact Lens–Induced Corneal Warpage: Irregular corneal shape changes caused by prolonged or improper contact lens wear that can mimic keratoconus topographically but typically resolves after discontinuing lens wear.
* Corneal Ectasia Post-Refractive Surgery: Progressive corneal thinning and bulging following procedures like LASIK, which can resemble keratoconus but is iatrogenic in origin.
* Posterior Keratoconus: A rare, non-progressive condition characterized by localized posterior corneal depression without anterior surface changes, differentiating it from typical keratoconus.
* Fuchs-Terrien Marginal Degeneration (FTMD): A peripheral corneal thinning disorder that can mimic ectatic changes but has distinct clinical and topographic features

## **Epidemiology**

* Prevalence: The prevalence of keratoconus varies widely worldwide, influenced by geographic location, ethnicity, diagnostic criteria, and detection methods. Reported prevalence ranges from as low as 0.0003% (0.3 per 100,000) in Russia to as high as 5% in some Middle Eastern populations, with other studies showing figures between 0.054% in the USA to 2.3% in Central India.
* Geographic Variation: Keratoconus is more common in hot, dry climates such as the Middle East, India, China, and Australia. Northern Europe, northern USA, Japan, Russia, and Finland report lower prevalence rates. Environmental factors like ultraviolet light exposure and ethnic background may contribute to these differences.
* Ethnicity: The disease is more prevalent among blacks and Latinos compared to whites, with odds ratios of 1.57 and 1.43 respectively.
* Age and Sex: Keratoconus usually manifests in the early 20s and progresses more rapidly in younger patients before stabilizing around 20 years after onset. It affects both males and females, though studies show mixed results regarding sex predominance.
* Genetics and Family History: Approximately 14% of keratoconus cases show evidence of genetic transmission. Individuals with a family history have a 15 to 67 times greater risk of developing the disease compared to those without such history.

## Risk Factors

* Eye Rubbing: A significant risk factor, with up to 70-80% of patients reporting frequent eye rubbing. This mechanical trauma leads to corneal inflammation and stromal collagen loss.
* Atopy and Allergy: The association with atopy is inconsistent; some studies show a positive correlation, while others do not. However, atopy may contribute indirectly by promoting eye rubbing.
* Environmental Factors: Exposure to ultraviolet light and oxidative stress in hot, dry climates may facilitate disease manifestation.
* Systemic Associations: Keratoconus is sometimes associated with systemic connective tissue disorders such as Down syndrome, Leber congenital amaurosis, and Ehlers-Danlos syndrome, though these are less common

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**KERATITIS**

*ALTERNATIVE NAMES: Another name for keratitis is “ulcerative keratitis”, which refers to an inflammation or irritation of the cornea that can lead to an ulcer.*

**DEFINITION / DESCRIPTION**

Bacterial [keratitis](https://www.aao.org/eye-health/diseases/what-is-keratitis) is an [eye infection](https://www.aao.org/eye-health/eye-infections) that affects the [cornea](https://www.aao.org/eye-health/anatomy/cornea-103). The cornea is the clear, dome-shaped window of the front of your eye.

Bacterial keratitis usually develops quickly. Left untreated it can cause blindness.

There are many different bacteria that cause keratitis. The two bacteria most commonly responsible for this type of infection in the U.S. are:

* *Staphylococcus aureus*
* *Pseudomonas aeruginosa*

Keratitis is an inflammation of the cornea — the clear, dome-shaped tissue on the front of your eye that covers the pupil and iris. Keratitis may or may not be associated with an infection. Noninfectious keratitis can be caused by a relatively minor injury, such as from wearing your contact lenses too long or getting a foreign body in the eye. Infectious keratitis can be caused by bacteria, viruses, fungi and parasites.

If you have eye redness or other symptoms of keratitis, make an appointment to see an eye specialist. With prompt attention, mild to moderate cases of keratitis can usually be effectively treated without loss of vision. If left untreated, or if an infection is severe, keratitis can lead to serious complications that may permanently damage your vision.

**CAUSES**

Causes of keratitis include:

* **Injury.** If any object scratches or injures the surface of your cornea, noninfectious keratitis may result. In addition, an injury may allow microorganisms to gain access to the damaged cornea, causing infectious keratitis.
* **Bacteria, fungi or parasites.** These organisms may live on the surface of a contact lens or contact lens carrying case. The cornea may become contaminated when the lens is in your eye, resulting in infectious keratitis. Poor contact lens hygiene or contact lens overwear can cause both noninfectious and infectious keratitis.
* **Viruses.** The herpes viruses — herpes simplex and herpes zoster — may cause keratitis.
* **Bacteria.** Staphylococcus, streptococcus and pseudomonas are common bacteria involved in keratitis.
* **Contaminated water.** Bacteria, fungi and parasites in water — particularly in oceans, rivers, lakes and hot tubs — can enter your eyes when you're swimming and result in keratitis. However, even if you're exposed to these organisms, a healthy cornea is unlikely to become infected unless there has been some previous breakdown of the corneal surface — for example, from wearing a contact lens too long.

**What Causes Bacterial Keratitis?**

The two main causes of [bacterial keratitis](https://www.aao.org/eye-health/diseases/what-is-bacterial-keratitis) are:

* [contact lens](https://www.aao.org/eye-health/glasses-contacts/contact-lens-102) use, especially extended-wear lenses
* eye injury

You risk infection from contact lenses if you wear them too long or do not take care of them correctly. Proper care of your contacts will lower your risk of developing a corneal infection.

**RISK FACTORS**

Factors that may increase your risk of keratitis include:

* **Contact lenses.** Wearing contact lenses — especially sleeping in the lenses —increases your risk of both infectious and noninfectious keratitis. The risk typically stems from wearing them longer than recommended, improper disinfection or wearing contact lenses while swimming.

Keratitis is more common in people who use extended-wear contacts, or wear contacts continuously, than in those who use daily wear contacts and take them out at night.

* **Reduced immunity.** If your immune system is weakened due to disease or medications, you're at higher risk of developing keratitis.
* **Corticosteroids.** Use of corticosteroid eye drops to treat an eye disorder can increase your risk of developing infectious keratitis or make existing keratitis worse.
* **Eye injury.** If one of your corneas has been damaged from an injury in the past, you may be more vulnerable to developing keratitis.

**SIGNS / SYMPTOMS**

Symptoms of keratitis include:

* Eye redness
* Eye pain
* Excess tears or other discharge from your eye
* Difficulty opening your eyelid because of pain or irritation
* Blurred vision
* Decreased vision
* Sensitivity to light, called photophobia
* A feeling that something is in your eye

**Bacterial Keratitis Symptoms**

Symptoms of bacterial keratitis may include:

* pain in the eye (often sudden)
* unusual eye redness
* reduced vision
* increased light sensitivity
* excessive tearing
* discharge from your eye

Call your ophthalmologist right away if you have any of these symptoms. This is especially important if they come on suddenly. If not treated, a bacterial keratitis eye infection can lead to blindness. [Treatment](https://www.aao.org/eye-health/diseases/bacterial-keratitis-treatment) must start right away to prevent vision loss.

**BACTERIAL KERATITIS DIAGNOSIS METHODS**

To diagnose bacterial keratitis, your ophthalmologist will discuss your symptoms with you. They may gently scrape the eye to take a small sample and test it for infection.

Bacterial keratitis is usually treated with antibiotic eye drops. Drops are usually put in frequently. Treatment may also involve steroid drops. You may need to return to your ophthalmologist several times.

If you and your ophthalmologist find and treat bacterial keratitis early, you may preserve your vision. In severe cases, decreased vision or blindness may be the result. This is also true if the infection affects the center of the cornea. Sometimes a cornea transplant or specialized contact lens is needed to restore vision.

**DIAGNOSIS METHOD**

Diagnosing keratitis typically involves the following:

* **Eye exam.** Although it may be uncomfortable to open your eyes for the exam, it's important to have your eye care provider examine your eyes.
* **Penlight exam.** Your eye doctor may examine your eye using a penlight, to check your pupil's reaction, size and other factors. A stain may be applied to the surface of your eye. Used with the light, this stain makes it easier to see damage to the surface of the cornea.
* **Slit-lamp exam.** Your eye care provider will examine your eyes with a special instrument called a slit lamp. It provides a bright source of light and magnification to detect the character and extent of keratitis, as well as the effect it may have on other structures of the eye.
* **Laboratory analysis.** Your eye care provider may take a sample of tears or some cells from your cornea for laboratory analysis to determine the cause of keratitis and to help develop a treatment plan for you.

**TREATMENT OPTIONS**

**Noninfectious keratitis**

Treatment of noninfectious keratitis varies depending on the severity. For example, with mild discomfort from a corneal scratch, artificial tear drops may be the only treatment. However, if keratitis is causing significant tearing and pain, topical eye medications may be necessary.

**Infectious keratitis**

Treatment of infectious keratitis varies, depending on the cause of the infection.

* **Bacterial keratitis.** Antibiotic eye drops are the primary treatment for bacterial keratitis. Depending on the severity of the infection, drop frequency can range from around four times a day to every 30 minutes, even during the night. Sometimes oral antibiotics are used as a supplement.
* **Fungal keratitis.** Keratitis caused by fungi typically requires antifungal eye drops and oral antifungal medication.
* **Viral keratitis.** If a virus is causing the infection, antiviral eye drops and oral antiviral medications may be effective. Other viruses need only supportive care such as artificial tear drops.
* Acanthamoeba keratitis. Keratitis caused by the parasite acanthamoeba can be difficult to treat. Antiparasitic eye drops are used, but some acanthamoeba infections are resistant to medication and can require treatment for several months. Severe cases of acanthamoeba keratitis may require a cornea transplant. Acanthamoeba causes keratitis, a sight-threatening, painful, and difficult-to-treat ocular infection that is linked to contact-lens wear. 1 The pathogen can also cause infections of the CNS and skin, which are associated with immunodeficiency, 2 although these types of infection are less common than keratitis.

If keratitis doesn't respond to medication, or if it causes permanent damage to the cornea that significantly impairs your vision, your eye care provider may recommend a cornea transplant.

**PREVENTION TIPS**

**Caring for your contact lenses**

If you wear contact lenses, proper use, cleaning and disinfecting can help prevent keratitis. Follow these tips:

* Choose daily wear contacts and take them out before going to sleep.
* Wash, rinse and dry your hands thoroughly before handling your contacts.
* Follow your eye care provider's recommendations for taking care of your lenses.
* Use only sterile products that are made specifically for contact lens care, and use lens care products made for the type of lenses you wear.
* Replace your contact lenses as recommended.
* Replace your contact lens case every 3 to 6 months.
* Discard the solution in the contact lens case each time you disinfect your lenses. Don't "top off" the old solution that's already in the case.
* Don't wear contact lenses when you go swimming.

**Preventing viral outbreaks**

Some forms of viral keratitis can't be eliminated. But the following steps may control viral keratitis occurrences:

* If you have a cold sore or a herpes blister, avoid touching your eyes, your eyelids and the skin around your eyes unless you've thoroughly washed your hands.
* Only use eye drops that have been prescribed by an eye doctor.
* Washing your hands frequently can reduce viral outbreaks.

**POSSIBLE COMPLICATION**

* Corneal scarring
* Corneal melt
* Corneal anesthesia
* Neurotrophic keratopathy
* Descemetocele
* Perforation
* Secondary glaucoma
* Neovascular glaucoma
* Iris Neovascularization
* Hyphema
* Hemorrhage
* Toxic iridocyclitis
* Subluxation of lens
* Anterior subcapsular cataract
* Corneal fistula
* Scleritis
* Retinal detachment
* Choroidal detachment
* Endophthalmitis
* Panophthalmitis
* Keratectasia
* Atrophic bulbi
* Autoevisceration
* Phthisis bulbi

**Surgical Complications**

* Wound leak
* Irregular trephination
* Small size graft
* Secondary glaucoma
* Flat anterior chamber
* Iridodialysis
* Pupillary block
* Expulsive choroidal hemorrhage
* Retinal detachment
* Choroidal detachment
* Vitreous hemorrhage

**Suture Related Complications**

* Vascularization
* Infection
* Loose sutures
* Wound leak
* Exposed knots

**WHEN TO SEE A DOCTOR**

If you notice any of the symptoms of keratitis, make an appointment to see an eye specialist right away. Delays in diagnosis and treatment of keratitis can lead to serious complications, including blindness.

**DIFFERENTIAL DIAGNOSIS**

**Infective**

* Fungal keratitis
* Pythium keratitis
* Viral keratitis
* Neurotrophic keratitis
* Neuroparalytic keratitis
* Interstitial keratitis
* Disciform keratitis
* Acanthamoeba keratitis
* Exposure keratopathy
* Chemical Injury
* Thermal keratitis
* Atopic keratoconjunctivitis
* Shield ulcer
* Rosacea keratitis

**Non- infective**

* Peripheral Ulcerative keratitis
* Marginal keratitis
* Mooren’s Ulcer
* Toxic keratitis
* Sterile inflammatory corneal infiltrate

**EPIDEMIOLOGY**

The incidence and prevalence of bacterial keratitis vary with the geographical location and local environmental and climatic risk factors. There is a vast disparity among populations in the incidence of bacterial keratitis in developed countries like the USA and Europe compared to developing countries like India, Nepal, Pakistan, and Bangladesh. This variation is because less industrialized nations have a significantly lower frequency of contact lens users. As a result, contact lens-related bacterial keratitis is also less.

In the USA, the reported incidence is 11/1 lakh users compared to Nepal with 799/1 lakh users. The annual incidence of microbial keratitis in the USA is 71,000 cases. The normal human eye rarely develops bacterial keratitis because the normal human cornea is resistant to infection. In a study by Ormerod et al. in North America, they described staphylococcal, *Pseudomonas*, and *Streptococcus* as the major causes of microbial keratitis. Neuman and Sjostrand, in their analysis, reported *Staphylococcus aureus* and *Staphylococcus epidermidis* as the most common gram-positive bacteria and Pseudomonas as the most common gram-negative bacteria.

The incidence of corneal ulcers has progressed from nearly zero in early 60 to 52% in the 1990s. Erie et al. also reported that the incidence of ulcerative keratitis rapidly progressed from 5.3 to 435% from 1950 to 1980. He concluded contact lens to be a significant contributor. The reported annual incidence of ulcerative keratitis in contact lens users is 4 to 21 per 10,000 extended wear and daily wear for soft contact lens wearers. The overall most common reported causes of bacterial keratitis are *Staphylococcus* and *Pseudomonas*. In studies from Italy and UK contact lens was found to be a more common predisposing risk factor for bacterial keratitis. Data from two hospital sets from Los Angeles reported gram-positive pathogens.

A two hospital analysis from Los Angeles, USA, reported coagulase-negative staphylococcus as the most common gram-positive pathogens and *Pseudomonas aeruginosa* as the most common gram-negative bacteria. Another analysis from the USA reported more prevalence of gram-negative organisms from the southern USA than the northern part of the country. A study from Texas reported gram-negative bacteria as the most common isolate.

Polymicrobial keratitis can also occur while analyzing culture. A study reported 43% of cultures having two or more bacterial microorganisms. *Staph. epidermidis* and *Fusarium* have been reported as the most common etiology for polymicrobial keratitis, with trauma being the most common risk factor. The Steroids for Corneal Ulcers Trial (SCUT) from South India reported 51.5% cases of Streptococcus *pneumoniae*, 22.75 cases of *Pseudomonas*, and 11.5% cases of *Nocardia*.

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**LOW VISION**

**Definition and description**

Low vision is a permanent visual impairment that you can’t correct with glasses, contacts or surgery. Most eye doctors define low vision as moderate to severe visual impairment — enough to inhibit your everyday activities, like driving and reading.

Terms like “partially sighted” and “legally blind” fall under the larger heading of low vision. But these terms also have more specific meanings. In the U.S., these refer to whether you qualify for special education or disability services for visual impairment.

Low vision may involve a variety of different types of visual impairment; not just nearsightedness or farsightedness, but also peripheral vision loss, a blind spot or blurred vision. You might have OK vision in some ways but still have low vision overall.

Low vision isn’t total blindness — some vision remains. Vision rehabilitation services help people with low vision make the most of what they’ve got. A specialist can help connect you with low vision aids and assistive technology to make your life easier.

#### **What qualifies as low vision on a visual acuity test?**

The standard visual acuity test in the U.S. is the Snellen eye chart. It measures the clarity of your vision at a distance of 20 feet from an object. If you have 20/20 vision, which is the average score, it means you can see an object clearly from 20 feet away.

If your score is 20/70, it means that your vision at 20 feet is like normal vision at 70 feet. The World Health Organization (WHO) classifies this score as moderate visual impairment or moderate low vision. A score of 20/200 or above qualifies as severe low vision.

But this isn’t the only score eye specialists consider. Other types of tests measure other aspects of your vision. For example, a visual field test measures your range of vision from side to side. Having a visual field of 20 degrees or less also qualifies as low vision.

Some people have low scores on vision tests, but they’re able to correct their impairment with glasses, contacts or surgery. If these treatments make your test scores go up, you don’t have low vision. Low vision is a condition you have even with your glasses on.

### **What causes low vision?**

Causes of low vision include acquired diseases, injuries and congenital (birth) defects.

Some of the most common causes include:

* Macular degeneration. This condition causes a gradual loss of central vision. You might develop a blurry or blind spot in your central visual field. It commonly affects people older than 50.
* Cataracts. Cataracts are another common age-related eye condition. A cataract is a cloud on your eye lens. Cataract surgery can often remove it, but not everyone is a candidate for surgery.
* Glaucoma. Glaucoma is a progressive condition that damages your optic nerve. It usually affects your peripheral vision and night vision first. Without treatment, it can do irreversible damage.
* Diabetes-related retinopathy. This condition causes blood vessels in your eye to leak fluids, which build pressure against your retina and damage it over time. It’s a complication of diabetes.
* Retinopathy of prematurity. Babies born preterm and treated with incubation can develop abnormal blood vessels in their retinas. In a minority of cases, these can cause lasting damage.
* Amblyopia (lazy eye). Amblyopia is a congenital defect that causes blurry vision in one eye. As your child’s brain relies more on the good eye and ignores the bad eye, that eye begins to drift.

Other causes include:

* Refractive errors. Refractive errors affect the shape of your eye, causing nearsightedness or farsightedness. They’re correctable, but if they go untreated, they’ll continue to worsen.
* Trachoma. Trachoma is a bacterial infection that can irritate and damage your eyes. It’s a leading cause of vision loss and blindness worldwide, especially in less developed countries.
* Strabismus (misaligned eyes). Strabismus is a problem with the muscles that control your eye movements and eye alignment. It usually begins to appear in infancy or early childhood.
* Nystagmus. This condition causes rapid, uncontrollable eye movements that may make it impossible to focus your vision. It can be present at birth or acquired later in adulthood.
* Retinitis pigmentosa. Retinitis pigmentosa describes a group of inherited eye diseases that prevent your retina from working properly. Different genetic mutations can cause it.
* Hypertensive retinopathy. Chronic high blood pressure can disrupt the blood flow to your organs, including your retinas. Vision loss may be your first symptom of blood flow problems.
* Retinal detachment. This painless but serious condition causes your retina to detach from the tissues that support it, losing its blood supply. It can cause sudden and permanent vision loss.
* Optic atrophy. Optic atrophy causes your optic nerve to gradually deteriorate (waste away). A variety of conditions can cause it, including injuries, infections and blood flow problems.
* Eye injuries. A traumatic injury to your eye can cause lasting damage, especially if it isn’t recognized or treated right away. Pay attention if you have vision problems after an injury.
* Brain injuries. A traumatic head injury or stroke may cause brain damage that affects your vision. You might notice vision problems along with symptoms like headaches and dizziness.
* Vitamin A deficiency. Vitamin A is crucial to your vision. If you don’t get enough from your diet, or your body can’t absorb enough, it can cause vision loss. Night blindness is the first symptom.
* Eye cancer. All forms of eye cancer are very rare, but they can cause vision loss. Treatments to remove eye cancer can also cause vision loss, either by damaging or removing parts of your eye.

### **What are the signs and symptoms of low vision?**

Low vision can manifest in different ways for different people.

You might have a loss of:

* Central vision: The ability to see what’s directly in front of you.
* Peripheral vision: The ability to see out of the sides of your eyes.
* Depth perception: The ability to judge the distance between objects.
* Contrast sensitivity: The ability to distinguish objects in the foreground from objects of the same shade in the background.
* Night vision: The ability to see at night or in low light.
* Glare resistance: The ability to function in bright light.

You might have difficulties with:

* Reading.
* Driving.
* Cooking.
* Classroom learning.
* Watching TV or videos.
* Using a computer.
* Recognizing people’s faces.
* Getting around, especially in unfamiliar places.

Signs of low vision in children might include:

* Frequently bumping into things.
* Holding objects very close to their face.
* Frequent squinting or blinking.
* Frequently shutting or covering one eye.
* Eyes that flutter or dance.
* Eyes that don’t point the same way.
* Pupils of different sizes.
* Pupils that look gray or white.

Side effects of permanent vision impairment can include:

* Reduced physical and social activity.
* Loss of independence or employment.
* Hallucinations (Charles Bonnet syndrome).
* Anxiety or depression.

**Diagnosis methods**

### **How is low vision diagnosed?**

An ophthalmologist can diagnose your visual impairment, using a variety of vision tests. They’ll tell you what the issue is, how severe it is and if it’s treatable or not. If it’s moderate to severe, irreversible and significantly impacts your life, they’ll diagnose low vision.

Receiving this diagnosis can be difficult, especially if you weren’t aware that you were at risk of irreversible vision loss. You’ll probably have a series of reactions, from shock and bewilderment to numbness, denial, anger or grief. These are all natural reactions.

## **Management and Treatment**

When you see an optometrist specializing in low vision, they’ll give you a special kind of exam called a low vision exam. They’ll begin by taking a complete history of your eye health, and then ask about how your condition is currently affecting your life.

They’ll ask you how low vision affects your:

* School or work.
* Reading and computer use.
* Driving.
* Functioning in the kitchen.
* Facial recognition.
* Ability to travel.
* Hobbies and leisure activities.
* Mood and social life.

The provider will also examine your eyes and vision to look for any changes in your condition. They’ll use special low vision test charts to evaluate your visual acuity.

They might also need to check your:

* Field of vision.
* Eye muscle function.
* Glare sensitivity.
* Contrast sensitivity.
* Night vision.
* Color vision.
* Depth perception.
* Reading ability.

Based on the results of your exam, a low vision specialist will design a personalized treatment plan that addresses your specific difficulties and needs. They’ll offer resources and recommendations to help you adapt and optimize your quality of life.

#### **Vision rehabilitation**

Treatment for low vision is called vision rehabilitation. The goal of the treatment is to maximize your vision as much as possible and otherwise help you live as independently as possible with the vision you have. This may involve a wide variety of resources.

Your plan might include:

##### **Low vision aids and devices**

You might benefit from:

* A prescription for glasses or contact lenses.
* Optical magnifiers or telescopes.
* Electronic magnifiers and screen readers.
* Large print and high contrast products.
* Voice-to-text and audio reading technology.
* Audible home devices.

##### **Practical training and support**

You might also benefit from:

* Occupational therapy to learn new ways to perform tasks.
* A mobility specialist to help you learn to get around.
* A rehabilitation instructor to teach you independent living skills.
* Special education or vocational services.
* Counseling or psychotherapy to maintain your mental health.
* Support groups that connect you with others living with low vision.

## **Prevention**

The best way to prevent permanent vision loss is to keep up with your regular eye exams and see your provider right away if you notice anything unusual. While not all causes of low vision are preventable, many are treatable if you catch them early enough.

## **Outlook / Prognosis**

Not everyone with low vision bothers with rehabilitation services, but most people could benefit a lot from them. From practical tools, tips and tricks to social and emotional support systems, there’s a wealth of resources out there to take advantage of.

Adapting to a disability isn’t easy, but it’s easier if you don’t try to do it alone. Remember, no matter what your challenges are, someone else out there has already faced them. Others have found solutions and ways of coping that they can pass on to you.

## **Living With**

You may be eligible for disability benefits, based on your specific diagnosis, your financial position and other factors. Different governments have different criteria for receiving benefits. Specialists on your visual rehabilitation team can help you apply.

**Differential diagnosis**

## Common Causes of Low Vision

* Age-related Macular Degeneration (AMD): The leading cause of central vision loss in older adults, involving degeneration of the macula.
* Cataract: Opacification of the lens causing blurred vision, often reversible with surgery.
* Diabetic Retinopathy: Microvascular damage in diabetic patients leading to retinal ischemia and vision loss.
* Glaucoma: Progressive optic neuropathy causing peripheral vision loss and eventual blindness if untreated.
* Retinal Vascular Occlusions: Including retinal vein and artery occlusions causing sudden vision loss.
* Vitreous Hemorrhage: Bleeding into the vitreous cavity that obscures vision.
* Corneal Ulcer and Dry Eye Syndrome: Surface diseases causing visual disturbance.
* Uveitis/Scleritis: Inflammatory conditions that can impair vision.
* Non Diabetic Myopic Lens Shift: Lens changes causing refractive errors.
* Wet AMD: Neovascular form causing rapid vision loss.
* Stroke and Migraine Aura: Neurological causes causing transient or permanent visual field defects.
* Pituitary Tumors: Can compress the optic chiasm leading to bitemporal hemianopia.

## Uncommon Causes

* Corneal Hydrops: Acute stromal edema in keratoconus causing sudden vision loss.
* Traumatic Vision Loss: Direct injury to the eye or visual pathways.
* Optic Neuritis: Inflammation of the optic nerve, often painful and associated with multiple sclerosis.
* Papilledema: Optic disc swelling due to increased intracranial pressure, causing transient visual obscurations.
* Leber Hereditary Optic Neuropathy (LHON): Mitochondrial genetic disorder causing bilateral central vision loss in young adults.
* Acute Angle-Closure Glaucoma: Sudden rise in intraocular pressure causing vision loss and pain.
* Retinal Detachment: Separation of retina from underlying tissue causing vision loss.
* Postoperative Endophthalmitis: Infection after eye surgery leading to vision impairment.
* Ischemic Optic Neuropathies: Arteritic and non-arteritic forms causing sudden vision loss.
* Transient Ischemic Attack (TIA): Temporary visual disturbances due to transient cerebral ischemia.
* Cancer-Associated Retinopathy: Paraneoplastic syndrome causing retinal degeneration.

## Additional Considerations

* Optic Nerve and Visual Pathway Lesions: Lesions at various points (optic nerve, chiasm, tract, radiations, occipital cortex) cause characteristic visual field defects such as hemianopias.
* Toxic and Nutritional Optic Neuropathies: Due to substances like alcohol, tobacco, or vitamin B12 deficiency.
* Retinitis Pigmentosa and Other Genetic Retinal Disorders: Progressive peripheral vision loss leading to low vision.
* Myopic Macular Degeneration and Optic Atrophy: Common in some populations as causes of low vision.

**Epidemiology data.**

* Global Prevalence: At least 2.2 billion people globally have some form of near or distance vision impairment, with about 1 billion cases considered moderate to severe vision impairment or blindness.
* Visual Impairment Cases: In 2019, there were approximately 437 million prevalent cases of visual impairment worldwide, representing a nearly 91.5% increase since 1990. The burden is higher in low socio-demographic index (SDI) regions and certain geographic areas such as Eastern Europe and Nepal.
* Age Distribution: Over 80% of people with moderate to severe visual impairment or blindness are aged 50 years or older. Childhood blindness prevalence is about ten times lower than in adults but remains a significant concern due to the lifelong impact.
* Gender Differences: Females have a higher risk of visual impairment than males, largely due to longer life expectancy and, in some regions, reduced access to eye care services.
* Regional Variation: Visual impairment prevalence varies widely by region, with higher rates reported in low- and middle-income countries, particularly in Africa, South-East Asia, and parts of the Eastern Mediterranean. For example, cataract is the leading cause of visual impairment in many regions, while uncorrected refractive error is also a major contributor.
* Causes: The major causes of low vision globally include cataract, uncorrected refractive errors, glaucoma, age-related macular degeneration, diabetic retinopathy, and corneal opacities. Glaucoma alone caused approximately 3.61 million cases of blindness and 4.14 million cases of moderate to severe visual impairment worldwide in 2020, accounting for about 8.4% of all blindness[6](https://www.nature.com/articles/s41433-024-02995-5)

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**MACULAR TELANGIECTASIA (MACTEL)**

**Definition and description**

Macular telangiectasia (MacTel) is a disease that affects the macula, causing central vision loss. It’s caused by abnormal blood vessels around the fovea. The fovea is the centre of the macula and is used for detailed central vision.

There are two main types of macular telangiectasia – type 1 and type 2. The blood vessel changes are different in each type.

## **Type 1 macular telangiectasia**

In Type 1 macular telangiectasia, the blood vessels are dilated (enlarged) and aneurysms (or bulging of blood vessels) form. This causes swelling and damage to the macula. It usually occurs in one eye only and is believed to be a variation of Coats disease. Coats disease is a rare retinal disorder that most often impacts young people.

## **Type 2 macular telangiectasia**

Type 2 is the most common form of macular telangiectasia. In Type 2, the blood vessels dilate (enlarge) and leak, causing swelling of the macula. Sometimes new blood vessels can form under the retina and also break or leak. Eventually, scarring over the macula can occur, severely affecting vision. This type of MacTel usually occurs in both eyes but can affect each eye differently.

Type 2 macular telangiectasia is typically diagnosed in people aged in their 40s and 50s. People with macular telangiectasia tend to have a higher prevalence of diabetes and high blood pressure.

## **Symptoms of MacTel**

In its early stages, people with macular telangiectasia may not have symptoms. As the disease progresses over 10 to 20 years, there may be blurriness and distortion of central vision depending on the severity of the disease. Side or peripheral vision is usually unaffected.

## **How is MacTel diagnosed?**

Signs of macular telangiectasia can be detected during a comprehensive eye examination by your optometrist or ophthalmologist. Your eye health professional may do further tests such as optical coherence tomography (OCT) and fluorescein angiography (FA) to confirm the diagnosis.

## **Treatment**

Unfortunately, there’s currently no effective treatment for macular telangiectasia.

In cases where new blood vessels have formed under the retina, your eye health professional may suggest injections of anti-vascular endothelial growth factor (anti-VEGF) into your eye to help prevent further vision loss.

## **Prognosis**

According to the MacTel Project, 60% of type 2 patients have a VA of 20/50 or better, with a mean VA of 20/40. Mac Tel can rarely lead to 20/200 or worse vision, especially when associated with full-thickness macular hole or retinal neovascularization associated with disciform scarring. However, about 79% of eyes with neovascularization and about 78% of eyes with macular hole have VA better than 20/200. Interestingly, eyes with VA 20/200 or worse (considered to be advanced disease) were reported to have pigmentary plaques, which are thought to be surrogate markers for photoreceptor degeneration.

## **Differential Diagnosis**

Differential diagnoses of retinal capillary telangiectasia include branch retinal vein occlusion, diabetic retinopathy, and radiation retinopathy. In cases of neovascularization, age-related macular degeneration should also be ruled out.

**Recent guidelines or updates**

# **Management**

There have been no randomized clinical studies in Mac Tel. In non-neovascular cases, laser therapy, intravitreal anti–vascular endothelial growth factor (VEGF) injections, and corticosteroids have all been shown to not be effective in controlling the disease. For neovascular cases, intravitreal anti-VEGF injections are the mainstay of treatment, although transpupillary therapy and photodynamic therapy have been used in the past.

Neuroprotective agents are currently under investigation for the treatment of Mac Tel type 2. A phase 2 trial found decreased ellipsoid zone loss, increased macular thickness, and stable reading speed with intraocular delivery of ciliary neurotrophic factor (CNTF). The phase 3 multicenter Renexus (NT-501) trial compared the safety and efficacy of an intravitreal ocular implant designed to deliver therapeutic doses of CNTF vs sham treatment.

Associated full-thickness macular holes may be treated with pars plana vitrectomy surgery, membrane peeling, and gas tamponade, but these procedures may have lower-than-average closure rates and corresponding postoperative VA.

**Epidemiology data.**

The epidemiology of macular telangiectasia type 2 (MacTel 2), a progressive retinal disease affecting central vision, shows the following key points:

* Prevalence Estimates: Population-based studies estimate the prevalence of MacTel 2 to range from approximately 0.06% to 0.12% in various populations. For example, in African populations (Kenya and Nigeria), the prevalence was about 0.06% (95% CI 0.02–0.14%). In predominantly white populations such as in the Beaver Dam Eye Study (USA), prevalence was around 0.1% among individuals aged 43–86 years[2](https://pmc.ncbi.nlm.nih.gov/articles/PMC2901890/). A Spanish study using multimodal imaging reported a prevalence of 0.12%.
* Age and Gender: MacTel 2 typically presents in the fifth to seventh decades of life, with mean ages around 60–62 years reported in studies. Females may be slightly more affected, as seen in the African cohort where 4 out of 5 affected individuals were female[1](https://pubmed.ncbi.nlm.nih.gov/22364548/).
* Incidence: A 20-year population-based study in Olmsted County, Minnesota, reported an incidence of approximately 0.7 cases per 100,000 persons per year (0.0007%).
* Underestimation: Prevalence figures are likely underestimated because most studies rely on color fundus photography without advanced imaging modalities such as fluorescein angiography or optical coherence tomography (OCT), which can detect earlier or subtler disease signs.
* Risk Factors: Associations with systemic conditions such as type 2 diabetes, hyperlipidemia, and hypertriglyceridemia have been observed, suggesting metabolic factors may contribute to disease risk. The role of smoking remains unclear, with some studies showing associations with certain retinal lesions but not directly with MacTel 2 diagnosis.
* Ethnic and Geographic Variation: The prevalence appears similar across different ethnic groups and geographic regions, although data are limited. African populations show prevalence comparable to predominantly white populations

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[Macular telangiectasia (MacTel) | Macular Disease Foundation Australia Macular Disease Foundation Australia](https://www.mdfoundation.com.au/about-macular-disease/other-macular-conditions/macular-telangiectasia/)

**Meesmann corneal dystrophy**

**Definition and description**

THIS is an eye disease that affects the cornea, which is the clear front covering of the eye. This condition is characterized by the formation of tiny round cysts in the outermost layer of the cornea, called the corneal epithelium. This part of the cornea acts as a barrier to help prevent foreign materials, such as dust and bacteria, from entering the eye.

In people with Meesmann corneal dystrophy, cysts can appear as early as the first year of life. They usually affect both eyes and increase in number over time. The cysts usually do not cause any symptoms until late adolescence or adulthood, when they start to break open (rupture) on the surface of the cornea and cause irritation. The resulting symptoms typically include increased sensitivity to light (photophobia), twitching of the eyelids (blepharospasm), increased tear production, the sensation of having a foreign object in the eye, and an inability to tolerate wearing contact lenses. Some affected individuals also have temporary episodes of blurred vision.

other names

* Corneal dystrophy, juvenile epithelial of Meesmann
* Corneal dystrophy, Meesmann epithelial
* Juvenile hereditary epithelial dystrophy
* MECD
* Meesman's corneal dystrophy
* Meesmann corneal epithelial dystrophy
* Meesmann epithelial corneal dystrophy

**Causes**

Meesmann corneal dystrophy can result from mutations in either the *KRT12* gene or the *KRT3* gene. These genes provide instructions for making proteins called keratin 12 and keratin 3, which are found in the corneal epithelium. The two proteins interact to form the structural framework of this layer of the cornea. Mutations in either the *KRT12* or *KRT3* gene weaken this framework, causing the corneal epithelium to become fragile and to develop the cysts that characterize the disorder. The cysts likely contain clumps of abnormal keratin proteins and other cellular debris. When the cysts rupture, they cause eye irritation and the other symptoms of Meesmann corneal dystrophy

## **Signs**

* Myriads of intraepithelial microcysts most prominently in the interpalpebral zone
* Round-to-oval shaped punctate opacities in the central corneal epithelium
* Recurrent corneal erosions
* Reduced corneal sensitivity
* Smooth corneal surface

## **Symptoms**

* Decreased visual acuity (usually minor)
* Foreign body sensation
* Photophobia
* Lacrimation
* Temporary episodes of blurred vision
* Eye stinging
* Blepharospasm

# **Diagnosis**

Although the presence of intraepithelial cysts becomes evident in the ﬁrst decade of life, the vast majority of patients tend to remain asymptomatic or experience mild symptoms that often go unnoticed until late adolescence or adulthood. Affected individuals are therefore often diagnosed incidentally during a routine ophthalmic examination. Overall, the disorder does not cause much discomfort or significant vision disturbances and symptoms are usually mild and include recurrent irritation and decrease in visual acuity. The first presented symptom is ocular irritation linked to recurrent corneal erosions such as photophobia, pain, foreign body sensation, and excessive tearing, particularly upon lid-opening during sleep or upon awakening. The second symptom that follows is blurred vision as a result of corneal clouding

## **Diagnostic Procedures**

* Slit-lamp examination in direct illumination: the lesions appear as a variety of different patterns (random, whirled, sectorial, interpalpebral or unilateral and sparing the perilimbal region) of multiple tiny grey dot-like opacities and tiny vesicles in the corneal epithelium, that tend to concentrate in the interpalpebral zone.Fluorescein usually fails to stain the microcysts, as they rarely open to the corneal surface.
* Slit-lamp examination in indirect illumination: the grey opacities appear as multiple solitary, round, and transparent cysts, rarely producing refractile lines due to a coalescence of several cysts, while the intervening cornea remains intact or slightly hazy.
* Retroillumination: the intraepithelial microcysts appear as refractile transparent dew drops and the vesicles present isolated. The characteristic bleb pattern of Meesmann corneal dystrophy can be best seen upon retroillumination
* Corneal elevation topography: marked irregularity of Placido rings
* In vivo confocal microscopy (IVCM): the microcysts present as hyporeflective, round-oval, or teardrop areas in the basal epithelial cell layer. The peculiar substance appears as reflective spots.
* Slit-Lamp Biomicroscopy and Photography: documentation of the biomicroscopic examination with photos is useful as the disease progresses.

# **Management**

## **General treatment**

Meesmann dystrophy is a genetic disorder that persists throughout life.MECD usually remains an entirely asymptomatic condition that requires no treatment. For patients who experience minor intermittent symptoms of ocular irritation, a conservative therapeutic approach largely aimed at symptoms is preferred. In rare cases of severe symptomatology or recurrent episodes of corneal erosions, surgical interventions may be required to prolong the reoccurrence of the disorder, and also lessen its severity.

## **Medical therapy**

Mild to moderate symptoms and MECD-associated chronic eye dryness can be treated by using lubricating eye drops during the daytime and gel/ointment at night, but the majority of patients do not usually require further treatment. Some physicians advocate the use of topical steroids to increase the stability of the epithelial basement membrane, while others prefer hypertonic saline, especially in gel or ointment form at night, which causes dehydration of the epithelium and promotes its attachment to the underlying layers. Patching, either conventional or using a bandage soft contact lens, can be beneficial as it minimizes the mechanical effect of lid movement on the irregular corneal epithelium.

## **Surgery**

A more severe phenotype of MECD, described by recurrent corneal erosions and secondary scarring, can complicate the clinical presentation of MECD and conservative therapy often proves to be unsuccessful. Surgical treatment options aim at the removal of the pathological corneal epithelium and include superficial corneal debridement, superficial keratectomy with or without mitomycin C, phototherapeutic keratectomy (PTK), lamellar keratoplasty, and penetrating keratoplasty. Although the dystrophy frequently recurs after any of these treatments, in some cases recurrence may be less severe, delayed, or not occur.

Superficial Mechanical Debridement of the epithelium can be useful in the management of RCE, but is usually followed by reappearance of symptoms.

Superficial Keratectomy is a safe technique for managing corneal erosions and although it may be curative, it is rarely warranted.

Phototherapeutic Keratectomy using a 193- nm excimer laser is the method of choice nowadays, replacing manual lamellar keratoplasty as the former treatment of choice. PTK can be repeated several times for the treatment of recurrent corneal erosions, thus postponing corneal transplantation (lamellar or even penetrating) for a long time. This treatment achieves the removal of superficial corneal opacities, smoothing of the corneal surface, as well as correcting irregular astigmatism, and finally promotes the adherence of the epithelium to the underlying layers.

Lamellar and Penetrating Keratoplasty are seldom indicated for the treatment of MECD.

Rarely, a more severe phenotype with corneal erosions and significant scarring accompanied by a marked decrease in visual acuity may require treatment by keratoplasty or corneal grafting. Although epithelium damage is likely to recur in grafts, the recurrent disease is often less severe. It is important to postpone grafting as long as possible because most patients require regrafting due to recurrences.

## **Surgical follow up**

Surgical procedures that aim to remove the abnormal corneal epithelium are not curative, as the dystrophy recurs in the regenerated epithelium, and therefore provide only temporary symptomatic relief. All surgical treatment options have a high risk of recurrence.

## **Investigative Therapies**

Limbal stem cell allo-transplantation may be an alternative to replace the affected epithelial stem cells, while inhibition of the abnormal gene using specific siRNA’s to silence the mutant allele has been evaluated *in vitro* and is a promising potential treatment strategy.

## **Prognosis**

The prognosis for these patients remains mostly favorable. In rare cases, vision can be permanently affected as a result of secondary scarring with vesicular rupture.

## **Differential diagnosis**

## Suspected cases of MECD need to be differentiated from other disorders of the corneal epithelium, such as:

## Vapor spray keratitis

## Epithelial basement membrane dystrophy or Cogan's microcystic corneal dystrophy

## Reis-Bücklers corneal dystrophy.

## Lisch epithelial corneal dystrophy (LECD)

## MECD and LECD appear to present clinical similarities, and the two disease entities need to be distinguished from each other. This can be achieved by studying the different patterns of inheritance, i.e. X-linked recessive versus autosomal dominant.

## **Epidemiology**

The prevalence of MECD is unknown, as an official registry of affected cases does not exist. It was first described in a large, multi-generational German family with more than 100 affected members and since then, the condition has been reported in individuals and families worldwide. Numerous cases have been identified in Denmark, Germany, the Netherlands, Northern Ireland, Switzerland, the USA, Japan, Saudi Arabia, Taiwan, and Poland

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[Meesmann Corneal Dystrophy - EyeWiki](https://eyewiki.org/Meesmann_Corneal_Dystrophy)

**MYDRIASIS**

**Definition and description**

If your pupils are dilated, the black center of your eyes (pupils) are larger than usual. Pupils are typically the same size in both eyes.

Pupils change in size to control how much light enters your eye. The colorful part of your eye (iris) controls the size of your pupil with tiny muscles. In bright light, your pupils will get smaller to prevent light from entering. In the dark, pupils get larger to allow more light in. These changes are called direct responses.

Pupils also shrink when you focus on a close object. This is called an accommodative response. If a pupil does not get smaller in bright light or expand in the dark, the pupil is not functioning normally.

### **mydriasis**

Mydriasis is when the pupil is dilated and doesn’t respond to light. Another term for mydriasis is “fixed pupil.”

A pupil’s normal size is 2 to 4 millimeters in bright light and 4 to 8 millimeters in dim light (dilated).

### **What do dilated pupils look like?**

Pupils are dilated when the center black portion of your eyes takes up more space than the colorful portion of your eyes (iris).

Pupils are supposed to dilate under normal circumstances due to light changes and emotional variables. Most of the time, dilated pupils will go back to normal size on their own. If pupils dilate suddenly, occur after a traumatic injury or cause headaches and confusion, seek medical attention immediately.

Yes, when one pupil is more dilated than the other it is called anisocoria. This common condition occurs when both pupils react normally to light but vary in size by more than half a millimeter. Anisocoria affects about 20% of the population.

**Causes**

### **What are the most common causes of dilated pupils?**

The most common causes of dilated pupils are:

· An eye exam (eye drops used to examine nerves and retina).

· A reaction to medication.

· A brain injury.

· The use of recreational drugs.

· Sexual arousal (increased production of oxytocin hormone).

· Adrenaline.

### **What recreational drugs cause dilated pupils?**

Certain recreational drugs can dilate pupils, including:

· Cocaine.

· Ecstasy (MDMA).

· LSD.

· Methamphetamines.

· Psychedelic mushrooms.

### **What medications cause my pupils to dilate?**

Certain over-the-counter and prescription medications can also lead to dilated pupils, including:

· Antidepressants.

· Antihistamines.

· Anti-nausea medications.

· Anti-seizure medications.

· Atropine.

· Botulinum toxin.

· Medications for Parkinson’s disease.

### **What conditions cause dilated pupils?**

Some medical conditions or injuries also dilate pupils, including:

· Adie's pupil (abnormal pupil response to light in one eye).

· Anisocoria (both pupils are different sizes).

· Eye injury.

· Head trauma.

· Microvascular cranial nerve palsy (blood flow to nerves is blocked).

· Migraine.

· Ocular migraine (one eye can experience a sudden array of changes, similar to migraine sufferers without the headache.)

**Diagnosis methods**

Diagnosing mydriasis involves a comprehensive assessment by a qualified doctor. The diagnostic process typically includes:

· **Physical Examination**: A thorough physical exam is conducted to assess pupil size and reactivity to light. The doctor will check for any visible signs of eye injury or abnormalities.

· **Medical History**: The doctor will review your medical history to identify potential causes of mydriasis, such as medication side effects, recent eye surgeries, or head injuries.

· **Visual Acuity Test**: A visual acuity test is performed to evaluate the quality of your vision and check for any impairments related to the dilated pupils.

· **Ocular Motility Test**: This test examines the muscles of the eye to assess their function and rule out any underlying neurological conditions that may be causing the mydriasis.

· **Blood Tests**: In some cases, blood tests may be ordered to rule out systemic conditions that can manifest with mydriasis as a symptom.

Based on the findings from these diagnostic procedures, the doctor can determine the underlying cause of the mydriasis and develop an appropriate treatment plan.

## **Treatment & Management**

The treatment for mydriasis depends on its underlying cause, with a focus on preserving eye function. If pupil dilation results from medication, doctors may recommend waiting for the drug’s effects to wear off instead of using additional medications to reverse it. However, in cases of brain or eye injuries, more aggressive treatments may be necessary. Severe trauma affecting the nerves or eye structures might require surgery to restore normal function. Some individuals may need to wear an eye patch during recovery to protect the affected eye. If mydriasis persists or worsens, medical intervention is crucial to prevent long-term complications. Here are some common treatment strategies:

· **Avoiding Direct Sunlight**: If mydriasis causes light sensitivity, it is important to avoid direct sunlight exposure and wear protective sunglasses when outdoors.

· **Medications**: In cases where mydriasis is caused by an allergic reaction to mydriatic agents, topical and systemic steroids or antiallergic medications may be prescribed to manage the symptoms.

· **Protective Eyewear**: Wearing opaque contact lenses or light-sensitive sunglasses can help alleviate discomfort and manage light sensitivity associated with mydriasis.

· **Prosthetic Contact Lenses**: In some cases, prosthetic contact lenses may be recommended to help reduce the amount of light entering the eye and improve comfort.

· **Surgery**: In rare instances where mydriasis is caused by an underlying condition that requires surgical intervention, such as a brain tumour or aneurysm, surgery may be necessary as part of the treatment plan.

It is important to follow the doctor's recommendations closely and attend follow-up appointments to monitor the progress of treatment.

## **Medication for Mydriasis**

Mydriasis is typically managed using topical medications that dilate the pupil. The commonly used medicines for mydriasis treatment include tropicamide, phenylephrine, cyclopentolate, and atropine. Tropicamide is used to produce mydriasis and cycloplegia for diagnostic procedures. It is an anticholinergic agent that relaxes the iris sphincter muscle to dilate the pupil. Phenylephrine is an adrenergic stimulant that acts on the iris dilator muscle. Cyclopentolate and atropine are also muscarinic antagonists, commonly used as mydriatics in various combinations. These medications can be administered in various formulations, including solutions, drops, and ophthalmic inserts. If you experience persistent or severe symptoms related to mydriasis, it is essential to consult a doctor for proper management.

### **Do dilated pupils hurt?**

Often, dilated pupils will cause symptoms based on how light reaches the eye. These include:

· Blurry vision.

· Headaches.

· Light sensitivity.

### **How are dilated pupils treated?**

Most of the time, dilated pupils will return to normal on their own without treatment, especially if they are the result of eye drops. For more serious cases of mydriasis, further treatment is required, including:

· Wearing sunglasses to reduce sunlight sensitivity (photochromic lenses, polarized lenses).

· Prosthetic contact lenses (to improve the appearance of eyes as the result of varying pupil size caused by trauma).

· Surgery (to repair eye damage from injury).

· Substance abuse rehabilitation.

### **How long will my pupils be dilated?**

If you received dilating eye drops from an ophthalmologist, your eyes could be dilated between four and 24 hours. The length of time is dependent on the type of drop used and how your body responds to it.

If pupil dilation is the side effect of a medication or drug, the duration may vary based on the type of drug and the dosage taken.

Pupil dilation that's a reaction to an emotional factor (adrenaline, attraction, stress) could have a shorter duration, and the pupil could return to normal size in as little as two to three minutes.

## **Prevention of Mydriasis**

Preventing mydriasis involves taking steps to avoid exposure to agents that can cause the condition. Here are some key preventive measures:

· **Practicing Proper Hand Hygiene**: Healthcare workers should follow strict hand hygiene protocols to minimize the risk of accidental exposure to anticholinergic agents that can cause mydriasis.

· **Avoiding Environmental Toxins**: Steer clear of contact with environmental toxins like Jimson weed (Datura stramonium) or belladonna alkaloids found in certain plants, as these can trigger pupil dilation.

· **Protective Eyewear**: If you're exposed to bright lights or certain chemicals, wearing sunglasses (particularly those with photochromic or polarised lenses) can help reduce light sensitivity.

· **Avoiding Certain Medications**: Use medications, such as antihistamines, muscle relaxants, and medicines used to treat glaucoma, as directed by a doctor if dilated pupils are observed.

· **Avoid Recreational Drugs**: Recreational drugs or excessive use of substances can lead to mydriasis. Avoiding these substances can help prevent pupil dilation.

By being aware of these potential triggers and taking appropriate precautions, the risk of developing mydriasis can be significantly reduced.

## **Complications**

In many cases, mydriasis, especially when triggered by certain medications or plant exposure, resolves on its own within a few hours or days.

During this time, individuals may experience increased sensitivity to light. To reduce discomfort, wearing sunglasses outdoors and avoiding bright lights is recommended. It is also advisable to limit driving until vision returns to normal.

If mydriasis occurs as a reaction to medication, the individual should avoid taking that drug in the future and consult their doctor about alternative options. Similarly, those affected by plant-related allergies, such as exposure to angel’s trumpet, should take precautions to prevent future contact.

Because dilated pupils are often associated with recreational drug use, individuals with mydriasis may face uncomfortable or misleading assumptions about their condition.

Mydriasis can lead to several complications, depending on the underlying cause and severity of the condition. Some of the potential complications include:

· **Blindness**: In severe cases, particularly those caused by brain injuries or neurological disorders, mydriasis can lead to vision loss or blindness.

· **Eye Irritation**: Dilated pupils can cause increased sensitivity to light, leading to eye irritation and discomfort.

· **Headaches**: Mydriasis can be accompanied by headaches, dizziness, and other symptoms due to increased light sensitivity.

· **Adie Syndrome**: This rare neurological disorder is characterised by a dilated pupil that does not respond to light, along with the absence of deep tendon reflexes. Adie syndrome can cause vision problems, eye pain, and sensitivity to light.

Prompt diagnosis and appropriate treatment of mydriasis can help prevent or manage these complications, ensuring better eye health and overall well-being.

## **When To Call the Doctor**

Although it is normal for dilation to occur based on changes in light, mydriasis could be a sign of an eye injury or problem within the brain, like a head injury, tumor or stroke. Contact your healthcare provider if you experience any of the following:

· Dizziness, headache or confusion (signs of a stroke).

· Sudden pupil dilation that isn’t caused by light change.

· Pupils don’t change when light changes.

· One pupil is larger than the other.

· Dilation persists for long periods of time.

**Differential diagnosis**

Physiological Causes

* Normal pupillary dilation in response to low light or emotional/sympathetic stimulation (e.g., excitement, concentration) .

## 2. Pharmacological Causes

* Use of mydriatic or anticholinergic drugs (e.g., atropine, scopolamine, tropicamide).
* Sympathomimetic drugs (e.g., phenylephrine, cocaine, amphetamines, ecstasy, LSD).
* Recreational drugs such as cocaine, methamphetamines, ecstasy, LSD, and marijuana .

## 3. Neurological Causes

* Oculomotor (Cranial Nerve III) palsy: Damage to the parasympathetic fibers causes unopposed sympathetic stimulation leading to mydriasis, often with ptosis and extraocular muscle weakness. Causes include aneurysms, brain herniation, tumors, stroke, trauma, or increased intracranial pressure .
* Benign Episodic Mydriasis: Transient unilateral mydriasis often associated with headaches, eye pain, and photophobia .
* Adie’s Tonic Pupil: Usually unilateral, with slow constriction to light and diminished deep tendon reflexes .
* Central nervous system disorders: Epilepsy, stroke, impending brain herniation, and other brain pathologies can cause mydriasis .
* Autonomic neuropathies: Parasympathetic dysfunction affecting pupil constriction .

## 4. Trauma

* Eye or head trauma damaging the iris sphincter muscle or the nerves controlling it, resulting in fixed dilated pupils .

## 5. Ocular Causes

* Acute angle-closure glaucoma causing mid-dilated pupil.
* Eye diseases such as glaucoma or high myopia.
* Damage to the iris sphincter muscle from surgery or inflammation .

## 6. Other Causes

* Increased oxytocin levels (e.g., during exercise or social interactions) may cause mild to moderate mydriasis .
* Plant poisoning (e.g., belladonna, Jimson weed) causing anticholinergic toxicity and pupil dilation .
* Systemic conditions such as infections or metabolic disorders affecting the autonomic nervous system.

**Epidemiology data**

Prevalence: In 2017, approximately 7.98 million cases of mydriasis were reported across seven major market nations, reflecting the widespread clinical relevance of this condition or its induced state. The United States has the highest prevalence of mydriasis among these countries.

* Market and Usage Trends: The mydriasis market, which includes drugs used to dilate pupils for diagnostic and surgical purposes, was valued at USD 641.5 million in 2024 and is projected to grow to USD 1,230.5 million by 2035, with a compound annual growth rate (CAGR) of about 6.1%. This growth is driven by an increasing prevalence of eye diseases such as cataracts, glaucoma, macular degeneration, diabetic retinopathy, and retinal diseases, which require pupil dilation for diagnosis and treatment.
* Demographics and Risk Factors: The rising aging population and greater awareness of eye health contribute to increased demand for mydriatic procedures. Additionally, the use of certain medications (e.g., antihistamines, anticholinergics) can induce mydriasis as a side effect, contributing to its epidemiology.
* Regional Distribution: Major markets include the United States, Germany, France, the United Kingdom, Italy, Spain, and Japan, with North America dominating due to advanced healthcare infrastructure and higher disposable incomes.
* Clinical Relevance: Mydriasis is a critical diagnostic tool in ophthalmology for better visualization of the retina and is essential in various ocular surgeries and laser treatments. Advances in drug formulations and delivery methods are improving safety and efficacy, which may influence epidemiological patterns by increasing procedure frequency

REFERENCE

<https://my.clevelandclinic.org/health/symptoms/22238-dilated-pupils>

**Nystagmus**

**Definition and description**

Nystagmus (pronounced “ni-STAG-muhs”) is a condition where your eyes make rapid, repetitive, uncontrolled movements. Your eyes may move in different directions:

* Side to side (horizontal nystagmus).
* Up and down (vertical nystagmus).
* In a circle (rotary or torsional nystagmus).

The movements can follow different patterns. Your eyes may:

* Drift in one direction and then jerk in the opposite direction to correct (jerk nystagmus).
* Drift back and forth in a steady, pendulum-like motion (pendular nystagmus).

These eye movements can cause problems with vision, depth perception, balance and coordination.

#### **Types of nystagmus**

Nystagmus affects both children and adults. There are two types: congenital or infantile (onset at birth or in the first few months of life) and acquired (onset after 6 months of age).

##### **Congenital or infantile nystagmus**

Babies born with nystagmus usually show symptoms between 6 weeks and 3 months of age. Sometimes, parents pass nystagmus on to their children, but the exact cause isn’t always clear. Children with congenital nystagmus often have it in both eyes. Their eyes usually move side to side. The main symptom is blurry vision.

##### **Acquired nystagmus**

Acquired nystagmus develops later in life and is more common in adults. Nystagmus may be a symptom of a medical condition affecting your brain, eyes or ears. Or, it may not be related to a condition at all. It may just be how your body works. Sometimes nystagmus results from alcohol and drug use. Adults with acquired nystagmus often describe their vision as shaky.

Spasmus nutans is a form of acquired nystagmus that affects children. It’s usually diagnosed between 6 months and 3 years old. This type of nystagmus usually improves without treatment between ages 2 and 8.

#### 

Researchers don’t know for sure how many people in the general population have nystagmus. Previous studies have reported that anywhere from 6 to 24 people out of every 10,000 have some type of nystagmus.

#### 

Nystagmus itself isn’t considered dangerous. But it may be associated with serious health conditions, especially those affecting your brain, such as stroke, brain tumor, toxicity, head trauma (injury) and inflammatory diseases.

### **What causes nystagmus?**

Your brain controls eye movement in conjunction with the structures in your ear, called the vestibular system. It automatically adjusts your eyes when you move your head so that the image you see remains in focus. In people with nystagmus, a problem prevents your brain, the vestibular system and your eyes from working together.

Nystagmus could indicate another eye problem, a neurological condition or a problem with the parts of your inner ear that control balance and coordination.

Nystagmus causes and risk factors include:

* Developmental problems with your brain or eye.
* Retina or optic nerve disorders.
* Inner ear disorders, such as benign paroxysmal positional vertigo (BPPV) and Ménière’s disease.
* Stroke.
* Brain tumor.
* Eye or head trauma (injury).
* Alcohol or drug use.
* Albinism (lack of pigmentation in the skin).
* Vision problems, including nearsightedness or astigmatism.
* Certain medications, such as antiseizure drugs.
* Diseases affecting your central nervous system, like multiple sclerosis (MS).
* Eye problems in babies, including strabismus (crossed eyes), focusing issues and cataracts.

Sometimes, there isn’t a clear cause. This is called idiopathic nystagmus.

### **What are the symptoms of nystagmus?**

The biggest sign of nystagmus is uncontrollable movement in your eyes. The symptoms of nystagmus depend on the condition causing it and include:

* Feeling as if your surroundings are moving (oscillopsia).
* Shaky or blurry vision.
* Balance problems.
* Light sensitivity or trouble seeing in the dark.
* Dizziness or feeling like you’re spinning (vertigo).

## **Diagnosis and Tests**

An eye care specialist called an ophthalmologist typically diagnoses nystagmus. They’ll perform an eye exam and ask about your symptoms. They’ll check for eye problems related to nystagmus, including strabismus, cataracts or issues with your retina or optic nerve. Other specialists, such as brain doctors (neurologists) and ear doctors (otorhinolaryngologists), can also diagnose nystagmus and test you for brain or inner ear conditions causing it.

#### **Tests to diagnose nystagmus**

You may need additional tests with different providers to learn what’s causing nystagmus. An ophthalmologist may perform tests to see if an eye disease is causing nystagmus. A neurologist may perform tests to see if a brain condition is causing nystagmus. An otorhinolaryngologist or audiologist may test to see if nystagmus relates to an inner ear condition.

Tests may include:

* A neurological exam.
* An ear exam.
* Eye movement recordings (such as electronystagmography and video-nystagmography).
* Imaging tests to capture pictures of your brain, such as CT scan (computed tomography scan) or MRI (magnetic resonance imaging).
* Genetic tests to identify inherited genetic mutations (errors in your DNA) associated with some forms of congenital nystagmus.

## **Management and Treatment**

The correction of nystagmus depends on the medical condition responsible for it.

Sometimes, treating the underlying condition can correct acquired nystagmus. For example, treating an inner ear condition causing nystagmus can improve symptoms like shaky vision or dizziness. In some conditions, your brain and vestibular system compensate for the damage, and nystagmus goes away or decreases over time.

Certain types of congenital nystagmus may disappear later in life. Other types can’t be cured completely, but proper treatment can manage symptoms.

### **What are the treatments for nystagmus?**

Your healthcare provider will recommend treatment based on what’s causing your nystagmus. They’ll also consider your health history and personal preferences.

#### **Glasses or contact lenses**

Clearer vision can help slow the rapid eye movements associated with nystagmus. Your provider may recommend eyeglasses or contact lenses to manage symptoms. You may need prism lenses, which limit how much your eyes must move to see clearly.

#### **Medications**

Some medications can reduce nystagmus symptoms in adults, such as gabapentin (antiseizure), baclofen (muscle relaxant) and onabotulinumtoxina (Botox®). Your healthcare provider will determine whether you would benefit from any medications.

#### **Eye muscle surgery**

In rare instances, your provider may recommend strabismus surgery. During this procedure, a surgeon repositions the muscles that move the eyes. This surgery doesn’t cure nystagmus, but it improves your eye movement. You won’t have to tilt or turn your head as much to see clearly.

#### **Vision correction surgery**

If you have nystagmus and are nearsighted, you may benefit from laser vision correction surgery — such as LASIK. Laser eye surgery doesn’t cure nystagmus, but it improves your vision. Improved vision can reduce your nystagmus symptoms.

## **Prevention**

Currently, there’s no way to prevent nystagmus. But you can reduce symptoms by treating the underlying cause.

## **Outlook / Prognosis**

Nystagmus can make everyday tasks more challenging. Sometimes, it limits the types of jobs and hobbies you can have.

Nystagmus rarely goes away completely, but it can improve over time. Your healthcare provider can help you find a treatment that works for you.

## **Living With**

### **When should I see my healthcare provider?**

Contact your healthcare provider immediately if you notice any changes in your vision or have difficulties with balance or coordination. Remember that nystagmus can be a symptom of serious health issues. Prompt diagnosis and treatment are essential.

If you’ve already been diagnosed with nystagmus, inform your provider if your symptoms worsen.

## **Epidemiology**

Mortality/morbidity

Mortality and morbidity are dependent upon etiology.

Age

All types of the acquired nystagmus described earlier, except spasmus nutans, can occur at any age. Onset of spasmus nutans is in infants aged 4-14 months with disappearance by age 5 years. Subclinical nystagmus, following the resolution of spasmus nutans, persists until at least 5 to 12 years of age.

Infantile nystagmus (juvenile nystagmus) is classified as onset before age 6 months and is discussed in a separate article.

**DIFFERENTIAL DIAGNOSIS**

## Physiological Nystagmus

* End-point nystagmus: Occurs at extreme gaze positions; fine jerk nystagmus.
* Optokinetic nystagmus: Induced by following moving objects; normal response.
* Voluntary nystagmus: Some individuals can induce nystagmus voluntarily.

## 2. Congenital Nystagmus

* Congenital Idiopathic Nystagmus (CIN): Presents before 2 months of age; horizontal, pendular or jerk; no ocular abnormalities; may be inherited (X-linked, autosomal).
* Sensory Nystagmus: Due to afferent visual system abnormalities limiting visual input (e.g., optic nerve hypoplasia, albinism, congenital cataracts, Leber congenital amaurosis).
* Latent Nystagmus: Appears only when one eye is occluded; associated with congenital strabismus.
* Associated with genetic conditions like aniridia, Noonan syndrome, and X-linked mutations (e.g., FRMD7 gene).

## 3. Acquired Nystagmus

* Vestibular Disorders: Benign paroxysmal positional vertigo (BPPV), Ménière’s disease, vestibular neuritis, superior canal dehiscence syndrome.
* Central Nervous System Disorders:
  + Stroke (especially brainstem or cerebellar)
  + Multiple sclerosis
  + Brain tumors (medulloblastoma, astrocytoma)
  + Trauma
  + Neurodegenerative diseases
  + Inflammatory conditions (encephalitis)
* Metabolic and Nutritional Disorders:
  + Wernicke’s encephalopathy (thiamine deficiency)
  + Other metabolic encephalopathies
* Toxic and Drug-Induced:
  + Alcohol intoxication
  + Anticonvulsants (phenytoin)
  + Benzodiazepines, barbiturates
  + Recreational drugs (amphetamines, MDMA, ketamine)
  + Lithium, SSRIs, pregabalin
* Ocular Causes:
  + Retinal disorders causing sensory nystagmus
  + High myopia or astigmatism
* Other Neurological Causes:
  + Oculogyric crises (distinguished from nystagmus)
  + Ocular bobbing
  + Trochlear nerve malfunction

## 4. Specific Types and Their Causes

* Downbeat Nystagmus: Often due to cerebellar or brainstem lesions, neurodegenerative diseases, or drug intoxication.
* Seesaw Nystagmus: Disconjugate eye movements, often from parasellar lesions affecting the optic chiasm.
* Pendular vs. Jerk Nystagmus: Helps localize lesion and etiology.

## 5. Other Causes

* Infections: COVID-19 associated neurological symptoms.
* Systemic Diseases: Canavan disease, Pelizaeus–Merzbacher disease, Whipple’s disease.
* Environmental: Miners’ nystagmus from prolonged low-light exposure.

REFERENCES

[Acquired Nystagmus: Background, Pathophysiology, Epidemiology](https://emedicine.medscape.com/article/1199177-overview#a6?form=fpf)

[Nystagmus: Definition, Causes, Testing & Treatment](https://my.clevelandclinic.org/health/diseases/22064-nystagmus)

**OCULAR TUMORS (EYE CANCER)**

These include benign and malignant growths affecting various eye structures.

ALTERNATIVE NAMES: Ocular tumors are also known as “eye neoplasm”, and sometimes referred to as “**Eye tumors”, or “eye neoplasm”.**

**Definition and description**

An eye tumor can be cancerous or noncancerous. About 3,490 new eye cancers are estimated to be diagnosed in 2023. Symptoms of eye tumors can include vision changes, pain, and changes in the shape and movement of the eye.

Eye cancer includes several rare types of cancers that begin in your eye, including your eyeball and the structures surrounding your eyeball. Eye cancer starts when cells multiply out of control and form a tumor. Tumors can be benign (noncancerous) or malignant (cancerous). Unlike benign tumors, malignant tumors can grow and the cancer can spread throughout your body.

Diagnosing and treating eye cancers early can often prevent the spread.

**What are the types of eye cancer?**

Healthcare providers categorize eye cancers based on where cancer starts, its location in your eye and the types of cells.

**Intraocular melanomas**

Intraocular melanoma arises from cells called melanocytes, the same type of cell involved in the most serious form of skin cancer (melanoma). Most eye cancers are melanomas. Most form in the middle part of your eye (uvea). They’re called uveal melanomas. They include:

* **Iris melanoma:** Forms in the colored part of your eye, or iris. It often produces a dark, growing spot that stands out against your iris. They tend to grow slowly.
* **Ciliary body melanoma:** Forms in the muscles that adjust your eyeball lens so you can see objects near and far. The ciliary body is behind your iris.
* **Choroidal melanoma:** Forms in the layer of your eyeball that keeps your retina (in the back of your eye) and the front of your eye nourished with blood. The choroid is the most common site for eye melanoma to form.

Melanomas sometimes form in the conjunctiva, the membrane that covers the front part of your eyeball. They’re called conjunctival melanomas. They’re incredibly rare. Like uveal melanomas, they tend to spread and are aggressive.

**Eyelid and orbital cancer**

Orbital and adnexal cancer form in the tissues close to your eyeball. Orbital cancers form in your orbit, or the tissues, muscles and nerves that move your eyeball. Adnexal cancer forms in supporting tissues, including your eyelids and tear glands. Healthcare providers classify them according to the type of cell that transforms into cancer.

Most are:

* **Squamous cell carcinoma:** Forms from the squamous cells in your top layer of skin.
* **Basal cell carcinoma:** Forms from the basal cells in your top layer of skin.
* **Rhabdomyosarcoma:** Forms inside muscle tissue.

**Retinoblastoma**

Retinoblastoma is a malignant tumor arising from the retina in the back of your eye. They’re most common in children under age five.

**Intraocular lymphoma**

Intraocular lymphoma is a rare form of B-cell lymphoma. It forms in white blood cells called lymphocytes. It’s most common in people who are older than 50 or who have weakened immune systems. Many people with this form of eye cancer also have primary central nervous system lymphoma (PCNSL). PCNSL is a cancer that may affect various parts of your central nervous system, including your brain, spinal cord and spinal fluid.

**How common is eye cancer?**

Eye cancer is extremely rare. Only about 3,400 people in the United States receive an eye cancer diagnosis each year. It’s more common for cancers to start in other parts of your body and spread to your eye. Because they don’t start in your eye, providers don’t consider these cancers eye cancer.

**What is the most common form of eye cancer?**

Intraocular melanomas are by far the most common form of eye cancer. Most start in the middle part of your eye in a structure called the choroid. Approximately 2,500 people in the United States receive this diagnosis each year.

**causes of eye cancer**

As with cancers in general, eye cancer occurs when cells begin to divide and multiply out of control, eventually forming a mass called a tumor. Pieces of the tumor can break off and spread to your lymph nodes and bloodstream. The cancer cells can travel to other parts of your body via your bloodstream and lymphatic system, causing new tumors to form in other organs. When this happens, healthcare providers say that your cancer has “spread” or “metastasized.” It’s a sign of a more advanced disease.

Scientists are still researching to understand what causes otherwise healthy cells to become cancer cells.

**Risk factors for eye cancer**

Researchers have identified several risk factors that may increase your likelihood of developing eye cancer:

* **Age:** Providers diagnose most eye cancers in people over 50. The exception is retinoblastoma, which affects children under age 5.
* **Skin color:** You’re more likely to get eye cancer if you’re white and if you have pale skin.
* **Eye color:** People with light eyes (blue, green) are more likely to get eye cancer than people with dark eyes (brown).
* **Inherited conditions:** Dysplastic nevus syndrome, an inherited condition that involves having several atypical-looking moles, can increase your risk of some eye cancers. BAP1 tumor predisposition syndrome can increase your risk of multiple cancers, including uveal melanoma.
* **Sunning and tanning:** It’s possible that exposure to UV rays from the sun or tanning beds can increase your risk of intraocular melanoma. Medical researchers need to conduct more research to know for sure.

**Signs and symptoms**

Many people with eye cancer don’t experience symptoms unless a tumor is growing in a location that interferes with how their eye works. Experiencing symptoms doesn’t mean you have eye cancer. Many benign (noncancerous) eye conditions share symptoms with eye cancer. See a healthcare provider to know for sure.

The most common symptom of eye cancer is painless vision loss. Other vision problems that may be signs of eye cancer include:

* Blurry vision.
* Vision loss (either partial or total).
* Seeing flashes of light, squiggly lines or spots (floaters).

Other signs and symptoms include:

* A bulging eye.
* Eye irritation that doesn’t improve.
* A dark spot in your iris that gets bigger.
* A growing lump on your eyelid or in your eyeball.
* Changes in your eyeball’s positioning in the socket and how it moves.

**What are the first signs of eye cancer?**

Most people don’t learn they have eye cancer until a healthcare provider, like an optometrist or ophthalmologist, notices something suspicious during an eye exam. For example, enlarged blood vessels in your eye or a dark spot may signal eye cancer or another eye condition. You’ll need tests to be sure.

**Diagnosis and Tests**

An eye disease specialist (ophthalmologist) or an ocular oncologist diagnoses eye cancer. They may perform a variety of procedures to rule out other, more common eye conditions before arriving at a cancer diagnosis.

**Eye exam**

During an eye exam, a healthcare provider examines your eye closely, looking for signs of cancer. They may look for dark spots and enlarged blood vessels. They may check to see if your eyeball is moving as it should. They may use special tools to see the structures in your eye more clearly.

* **Ophthalmoscope:** An ophthalmoscope is a handheld instrument that contains a light and multiple lenses. It allows healthcare providers to view structures in the back of your eyeball, like your retina.
* **Slit lamp:** A slit lamp is a device that sits on a platform and table. It uses a light source and special lenses, like a microscope, to see detailed views of the front and back of your eyeball.

**Imaging**

The results of imaging procedures and the information from your eye exam are often enough to diagnose eye cancer. Common imaging procedures include:

* **Ultrasound:** Ultrasounds use sound waves to create images of the inside of your eyeball. Ultrasounds show how big a tumor is and its location. They’re especially useful in diagnosing intraocular melanomas.
* **Fluorescein angiography:** During this procedure, a healthcare provider injects dye into your bloodstream that makes your blood vessels show up more clearly during imaging. Once the dye has had time to reach the blood vessels in your eyeball, your provider uses a special camera that shows how blood is flowing in your eyeball.

You may need additional imaging procedures if your provider suspects the cancer’s spread. Imaging procedures that can show if cancer’s spread to parts outside your eye include:

* Ultrasound.
* Chest X-ray.
* CT scan.
* MRI.
* PET scan.

**Biopsy**

During a biopsy, a healthcare provider removes a sample of tissue from the tumor and tests it for cancer cells. Healthcare providers can identify most eye cancers with a physical exam and imaging. Still, a biopsy can provide information about the makeup of your cancer cells, including genetic mutations (changes) that make them unique. Your provider can use this information to determine characteristics about your cancer, like how aggressive it is. It can also show if you’re eligible for certain treatments.

* **Fine needle aspiration biopsy:** A tiny needle removes a sample of fluid from your eye to test for cancer cells.
* **Incisional biopsy:** A provider removes part of the tumor and tests the tissue for cancer cells.
* **Excisional biopsy:** A provider removes the entire tumor and tests the tissue for cancer cells.

**How is eye cancer staged?**

Cancer staging helps providers determine how advanced cancer is. They use this information to plan treatments and gauge your prognosis, or the likely outcome of your condition.

There are two common staging systems for eye cancer:

Providers stage cancer by assessing various factors.

* **T:** The tumor’s size and whether it’s grown into nearby parts of your eye.
* **N:** Whether the tumor’s spread to the lymph nodes in your ear and neck.
* **M:** Whether the cancer’s spread to other organs (usually your liver).

They consider this information together to assign eye cancer a stage between I and IV, with I being the least advanced and IV meaning the cancer’s more advanced.

**Collaborative Ocular Melanoma Study (COMS) staging system**

Another common system stages cancer based on tumor size. Size influences the type of treatments that will likely work best. Measurements are in millimeters (mm).

* **Small:** Between 1 mm and 2.5 mm in height and 5 mm to 16 mm in width.
* **Medium:** Between 2.5 mm and 10 mm in height and 16 mm or less in width.
* **Large:** Bigger than 10 millimeters in height and 16 millimeters in width.

**What additional tests will be done to diagnose eye cancer?**

Your provider may recommend liver imaging scans if they suspect your cancer’s spread. Your liver is the most common place for eye cancer to spread outside of your eye.

**Management and Treatment**

For slow-growing tumors or if the diagnosis isn’t certain, your provider may recommend monitoring your condition and delaying treatment — especially if treatment risks outweigh the benefits. For example, you may want to delay treatment if treating an area could cause vision loss.

**Radiation therapy**

Radiation therapy is one of the most common treatments for eye cancer.

* **Brachytherapy:** Brachytherapy, or internal radiation therapy, is the most common treatment for eye melanomas. For treatment, your provider will implant a tiny disc near the tumor that releases radiation to kill cancer cells.
* **External beam radiation therapy (EBRT):** With EBRT, a machine that never touches your body directs radiation toward a tumor. Techniques include stereotactic surgery, which directs high doses of radiation toward your tumor in one treatment session. Proton beam radiation therapy is another option. This is a newer form of radiation therapy that delivers precise, high doses of radiation toward tumors. As the equipment is very expensive to maintain, it isn’t available everywhere.

**Surgery**

Surgery is a common treatment option, especially for small tumors that haven’t spread beyond your eyeball. Procedures include:

* **Iridectomy:** Removes part of your iris. Providers commonly use this procedure when treating small melanomas.
* **Iridocyclectomy:** Removes part of your iris and ciliary body. Providers commonly use this procedure when treating small melanomas.
* **Transscleral resection:** Removes melanomas in your choroid or ciliary body.
* **Enucleation:** Removes your eyeball. You may need this surgery for large tumors or when there’s no way to preserve your vision with treatment. Afterward, you’ll get an artificial eyeball that matches your remaining eye. Your healthcare team will work with you closely to fit you with a replacement that looks and moves like your eye.
* **Orbital exenteration:** Removes your entire eyeball and some of the surrounding tissue. Your provider may recommend this procedure if the cancer’s spread into structures surrounding your eyeball. As with enucleation, you’ll get an artificial eyeball implant afterward.

**Laser therapy**

Laser therapy uses heat to destroy eye cancer. The most common type is transpupillary thermotherapy (TTT). During the procedure, infrared light delivers concentrated heat toward the tumor, destroying cancer cells. Providers may use this on its own or after brachytherapy to prevent cancer from returning (recurring).

**Immunotherapy**

Immunotherapy treatments help your immune system identify and destroy cancer cells more effectively. In certain instances, providers use the immunotherapy drug tebentafusp to treat uveal melanoma. Immunotherapy is a common treatment for cancer that’s spread or that providers can’t surgically remove.

**Targeted therapy**

Targeted therapy drugs target specific weaknesses in cancer cells, destroying them. You may be eligible for targeted therapy treatments if cancer cells contain a *BRAF* gene mutation (change). Currently, this mutation is more common in skin melanomas, but this treatment may be beneficial in people with eye melanomas, too.

**Chemotherapy**

Chemotherapy isn’t a common treatment for eye cancer, but your healthcare provider may recommend it if your cancer hasn’t responded to other treatments or if it spreads to other areas.

**What are possible treatment side effects?**

Side effects depend on the type of treatment your provider recommends. As these treatments target your eye, it’s possible that you’ll experience vision changes. One of the most significant risks is partial or complete vision loss. These risks depend on multiple factors that you should discuss with your provider.

**Prevention**

There’s no way to prevent eye cancer. Still, you can improve your prognosis by getting screened if you know that you’re in a high-risk group for getting eye cancer. For example, you may consider regular exams if you have BAP1 tumor predisposition syndrome. If you have a family history of retinoblastoma and have a child, it’s a good idea to get them regular eye exams to screen for cancer.

**Outlook / Prognosis**

Your prognosis, or likely treatment outcome, depends on many factors, including the tumor’s size, location and how much it’s spread. For example, brachytherapy eliminates 95% of small and medium intraocular melanomas. Eye cancer may not be curable. However, its growth within your eyeball can be contained.

Ask your healthcare provider about your prognosis based on your specific type of eye cancer.

**What is the survival rate of eye cancer?**

Survival rates communicate information about how many people with a certain cancer diagnosis are alive five years after their diagnosis when compared to people without the same diagnosis. The survival rates for the most common form of eye cancer, intraocular melanoma, are excellent when a provider diagnoses and treats the cancer when it’s still in your eyeball. The survival rates aren’t as good if the cancer’s spread to other organs.

Fortunately, providers diagnose and treat most cancers before they’ve metastasized.

**Epidemiology of ocular surface and orbital tumors**

The clinical diagnosis of ocular surface tumors can be challenging due to their similar signs and symptoms. Therefore, collecting more cases and differentiating between pathological types and epidemiological changes is crucial to enhance the identification of these tumors. Benign tumors are frequently found in the eyelid area, with the conjunctiva, orbit, cornea, and limbus following suit in occurrence.

We aimed to elucidate the reason behind the limited diversity observed in the occurrence of eye tumor sites. The high incidence of eyelid tumors could be attributed to the intricate nature of the eyelid’s placement in the outermost layer of the eye’s structure and its constant exposure to the external environment, which may facilitate the growth of tumors.

Previous research has indicated that ocular tumors in children are linked to genetic factors and an abnormality in embryonic growth processes. Eyelid and ocular surface tumors are the most common types of ocular tumors in epidemiological studies. Most eyelid and ocular surface tumors are benign; a few are malignant. Intraocular and orbital tumors are mainly malignant. Calcified epithelioma is a common benign tumor in children’s eyelids. The cortical differentiation of the epidermis forms it and is a benign tumor in the deep skin. The most common malignant tumors on their eyelids and ocular surfaces in individuals aged 65 and above are squamous, basal, and melanoma. These tumors are frequently found in the eyelid and orbit.

Cavernous hemangioma is the most frequently occurring orbital site, typically affecting young individuals and presenting with slow proptosis as the main symptom. The histopathological imaging features of cavernous angioma involve irregular expansion of vascular spaces, partial capillary hyperplasia, and infiltration of interstitial plasma cells. Adult orbital malignancies primarily comprise lymphoma, a proliferative disease of immune cell tumors with multiple pathological types. Most cases are diagnosed as mucosa-associated lymphoid tissue (MALT) lymphomas, considered low-grade malignant lymphomas. Clinical symptoms and imaging studies can be challenging to differentiate from other conditions, such as specific inflammation, leading to misdiagnosis. Accurate diagnosis requires pathological examination and immunohistochemistry.

Tumors in the eyelids are among the most common ocular tumors, often caused by external contact or stimulation. Unfortunately, many of these tumors can be difficult to distinguish from inflammatory lesions, and malignant tumors can even metastasize and damage ocular health. Recent studies have shown that the number of patients with eyelid tumors is increasing each year, particularly for benign tumors like seborrheic keratosis and xanthoma. This could be due to changes in people’s lifestyles, cultures, and material consumption levels, as well as an increase in metabolic disorders among the population. For example, xanthoma, a systemic disease that often manifests as skin lesions, frequently appears on the eyelids. In malignant tumors, distinguishing between melanocytes can be challenging, and most proliferative lesions may develop into malignant melanoma if specific conditions are met. Basal epithelial cells may also have the ability to produce melanin, which can lead to malignancy. BCC is a common ocular surface malignancy occurring almost on the eyelids. Usually, nodules with pearl margins, central ulceration, and telangiectasia vessels. Risk factors include old age, sun exposure, and immunosuppression, with high recurrence rates. Moreover, sebaceous carcinoma (SCC) often affects older people, usually from the eyelid glands, with early symptoms of mild and insidious development.

Studies have shown that ocular surface microbes, temperature, and ultraviolet (UV) light can impact the conjunctival microenvironment. These Studies have shown that ocular surface microbes, temperature, and UV light can impact the conjunctival microenvironment. C.L. Shields et al. investigated 5002 patients and found that identifying benign and malignant conjunctival tumors had valuable clinical features. However, experience based on clinical work still requires a histopathological diagnosis. Goblet cells and lymphatic vessels are widespread in the conjunctiva, and irritation or blockage creates cysts. Conjunctival cysts are divided into dermoid and epidermoid, which can occur after surgery due to the invagination of conjunctival epithelial cells, and both capsule walls can be coated with laminated squamous epithelium. Unlike congenital cysts, goblet cells are standard cells of the cystic wall, where, in a few cases, the wall is composed entirely of goblet cells, and the cavity is filled with jelly samples. Sebaceous adenocarcinoma is most found in elderly patients and those with conjunctival issues. Most cases occur in the area surrounding the eye, where the sebaceous glands are most abundant

The cornea plays a crucial role in the path of light, and maintaining its structure and function is essential for normal vision and refraction. The development of corneal tumors can result in significant visual impairment or even blindness. Malignant tumors in the cornea are also at risk of metastasizing. Distinguishing between corneal neoplasia and hyperplastic tumor-like lesions poses a clinical challenge, with dermoid tumors being the most frequent benign tumors found in the cornea and limbus.

The limbus is the boundary between the clear cornea and the white sclera, covering the conjunctiva. Research has shown that limbal stem cells can experience malignant degeneration like other tissues in the limbus; recurrent epithelial defects may be related to the limbal stem cell defect. Due to the limbal pathological changes’ unique anatomical structure and location, there are many misunderstood conditions, including Mooren ulcers and corneal dystrophy.

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**OPTIC ATROPHY**

**ALTERNATIVE NAMES: Optic neuropathy**

**Definition and description**

Optic atrophy is a condition that affects the cells of your optic nerve, which carries impulses from your eye to your brain. “Atrophy” means to waste away or deteriorate. Optic nerve atrophy is another name for optic atrophy, and it’s a serious condition.

Atrophy of the optic nerve is a condition that happens because of long-term damage to optic nerve fibers from many different causes. Optic atrophy can cause irreversible issues with vision, including blindness.

### **What causes optic atrophy?**

Nerve fibers that transmit impulses to your brain make up your optic nerve. In the case of optic atrophy, something is interfering with your optic nerve’s ability to transmit these impulses. Many factors can cause this interference, including:

* Lack of proper blood flow (vascular/ischemia): This is the most common cause of optic nerve atrophy.
* Conditions that you’re born with or inherit (congenital): One condition, Leber hereditary optic neuropathy, causes you to lose vision in one eye first and then the other.
* Pressure from outside (extrinsic compression) and inside the nerve (intrinsic compression): The pressure can come from tumors. Glaucoma can damage your optic nerve and sometimes includes high eye pressure (intraocular pressure).
* Damage from inflammation, either from other diseases or swelling in the optic nerve itself: One cause is optic neuritis, which is inflammation of your optic nerve. Another is hydrocephalus, or fluid collection in your brain.
* Damage from infections: These infections can include diseases like syphilis and measles.
* Trauma (eye injury): Eye injuries can happen in many ways, including industrial or car accidents, sports or fighting.
* Damage from diseases of the retina: Retinal diseases include diabetes-related retinopathy and retinal vein occlusion.
* Damage from toxins, nutritional deficits or medications:Optic nerve atrophy from these causes usually affects both eyes.

#### **Is optic atrophy contagious?**

You can’t catch optic atrophy from anyone, but many types of infections can cause the condition. These infections include:

* Syphilis.
* Measles.
* Tuberculosis.
* Mumps.
* Chickenpox.
* Lyme disease.
* Aspergillosis.
* Cryptococcosis.

### **Symptoms of optic atrophy**

Optic atrophy symptoms relate to changes in vision, including:

* Blurred vision or a reduction in sharpness of vision.
* Difficulties with peripheral vision.
* Difficulties with color vision.

**Diagnosis methods**

### **How is optic atrophy diagnosed?**

It’s important to see an eye care specialist if you have any vision issues. They’ll begin by asking questions about your symptoms and your medical history. They may ask about what you eat, drink and what drugs or medications you take, including supplements. Then, they’ll do an eye exam.

#### **What tests will be done to diagnose optic atrophy?**

During testing, your provider will use an ophthalmoscope to look at your eyes. With optic atrophy, there are cell changes that providers can see, along with a paleness to your optic disc. The lack of color is related to blood flow changes in optic atrophy.

Your provider, who may also be looking for things like multiple sclerosis or tumors, may rely on other tests like these:

* Optical coherence tomography.
* Visual field tests.
* Magnetic resonance imaging (MRI) scans.
* Fluorescein angiography.
* Ultrasound.
* Blood tests.

## **Management and Treatment**

There’s no real cure or treatment for optic atrophy. This is why it’s so important to have regular eye exams and to see your healthcare provider right away for vision changes.

Your provider will need to treat the cause of the optic atrophy to stop the condition from getting worse. For instance, they may need to remove a tumor or remove fluid from your brain and spinal cord.

## **Prevention**

In most cases, you can’t prevent optic atrophy. You may be able to decrease your risk in some cases. For instance, you can:

* Wear safety goggles to prevent eye injuries.
* Get necessary vaccines to prevent infections when possible.
* Practice safe sex to avoid sexually transmitted infections.

## **Outlook / Prognosis**

### **What can I expect if I have optic atrophy?**

The outlook for people with optic atrophy depends on what’s causing the issue. If the cause is optic neuritis, you can usually count on eventually getting your vision back when the inflammation goes away. If the cause is some other optic neuropathy, your vision might not improve.

If you have glaucoma, early diagnosis can lead to successful treatment and slower growth of optic atrophy. Early diagnosis of a tumor can lead to prompt treatment and better outcomes. This can relieve the pressure on your optic nerve and prevent further damage.

If your sight worsens, ask your provider about ways to cope with low vision. There are low-vision aids like special lenses and magnifying glasses. There are also non-optical materials, like text-reading software.

## **Living With**

### **When should I see my healthcare provider about optic atrophy?**

You should see a provider for regular eye exams, especially if you have any type of eye-related conditions. You should consult a provider if you have any changes in your vision, like blurry vision, or difficulties with color vision or with peripheral vision.

If you have a sudden loss of vision or extreme eye pain, call your provider or get immediate medical help. These can be symptoms of serious eye conditions.

**Possible complications**

**What are the complications of optic atrophy?**

Optic atrophy can’t be reversed. The major complication is loss of vision, or blindness.

**Recent guidelines or updates**

**Promising Treatments and Future Outlook**

In recent years, there have been significant advancements in the field of optic atrophy research, leading to the development of promising treatments that offer hope for individuals with this condition.

One of the most exciting areas of research is gene therapy. Scientists are exploring the use of gene therapy to target and repair the specific genetic mutations that cause optic atrophy. By delivering healthy copies of the affected genes into the cells of the optic nerve, it is hoped that vision loss can be halted or even reversed. While still in the early stages of development, gene therapy holds great promise for the future of optic atrophy treatment.

Another potential treatment avenue is stem cell therapy. Stem cells have the unique ability to differentiate into various cell types, including those found in the optic nerve. Researchers are investigating the use of stem cells to replace damaged or lost cells in the optic nerve, with the goal of restoring vision. Although this approach is still experimental, early studies have shown promising results.

Additionally, neuroprotective therapies are being explored to prevent further damage to the optic nerve. These therapies aim to protect the remaining healthy cells and promote their survival. Various drugs and compounds are being studied for their potential neuroprotective effects, including antioxidants, growth factors, and anti-inflammatory agents.

Furthermore, advances in prosthetic devices and visual aids are improving the quality of life for individuals with optic atrophy. Retinal implants, for example, can bypass the damaged cells in the retina and directly stimulate the remaining healthy cells, allowing for the perception of light and shapes. Other assistive technologies, such as magnifying glasses, screen readers, and voice-activated devices, are also helping individuals with visual impairments to navigate their daily lives.

While these treatments and technologies show great promise, it is important to note that they are still in the research and development phase. Clinical trials and further studies are needed to determine their safety, efficacy, and long-term effects. However, the progress being made in optic atrophy research provides hope for a brighter future for patients with this condition.

**Optic Nerve Regeneration**

Optic nerve regeneration is a field of research that holds great promise for restoring vision in individuals with optic atrophy. The optic nerve is responsible for transmitting visual information from the eye to the brain, and any damage to this nerve can result in vision loss. Traditional belief held that the optic nerve cannot regenerate once it is damaged, but recent advancements in regenerative medicine and innovative techniques have challenged this notion.

One approach being explored for optic nerve regeneration is the use of stem cells. Stem cells have the unique ability to differentiate into various cell types, including nerve cells. Researchers are investigating the potential of using stem cells to replace damaged or lost nerve cells in the optic nerve. This could potentially restore the transmission of visual signals and improve vision.

Another promising avenue for optic nerve regeneration is the use of gene therapy. Gene therapy involves introducing specific genes into cells to correct genetic abnormalities or promote the growth of new cells. Scientists are studying the possibility of using gene therapy to stimulate the regeneration of optic nerve cells and enhance their functionality.

In addition to stem cells and gene therapy, researchers are also exploring the role of neurotrophic factors in optic nerve regeneration. Neurotrophic factors are proteins that support the growth and survival of nerve cells. By delivering these factors directly to the damaged optic nerve, scientists aim to promote the regeneration of nerve fibers and improve visual outcomes.

While optic nerve regeneration is still in the early stages of research, these advancements offer hope for the development of effective treatments in the future. Continued studies and clinical trials are necessary to further understand the mechanisms involved in optic nerve regeneration and to determine the safety and efficacy of these approaches. With further progress, it is possible that optic nerve regeneration could become a viable treatment option for individuals with optic atrophy, ultimately restoring their vision and improving their quality of life.

**Artificial Vision and Prosthetics**

Artificial vision and prosthetics have emerged as promising solutions for individuals with optic atrophy. These innovative technologies aim to restore limited vision and improve the quality of life for those affected by this condition.

Visual prostheses, also known as bionic eyes, are designed to bypass damaged optic nerves and stimulate the remaining functional parts of the visual system. These devices consist of an external camera that captures visual information and converts it into electrical signals. These signals are then transmitted to an implanted electrode array, which stimulates the remaining healthy retinal cells or the visual cortex directly.

One of the most well-known visual prostheses is the Argus II Retinal Prosthesis System. This device has been approved by the U.S. Food and Drug Administration (FDA) for use in individuals with severe to profound retinitis pigmentosa, a condition that can lead to optic atrophy. The Argus II system includes a small camera mounted on a pair of glasses, a portable video processing unit, and an implanted retinal prosthesis. It works by converting visual information into electrical signals that are transmitted wirelessly to the retinal prosthesis, which then stimulates the remaining retinal cells to create visual perceptions.

While visual prostheses have shown promising results in restoring limited vision, it is important to note that the quality and clarity of the restored vision may vary among individuals. The level of visual improvement depends on various factors, including the severity of optic atrophy and the overall health of the visual system.

In addition to visual prostheses, researchers are also exploring other innovative approaches to artificial vision, such as optogenetics. Optogenetics involves genetically modifying retinal cells to make them sensitive to light and then using light stimulation to activate these cells and generate visual signals. Although still in the experimental stage, optogenetics holds great potential for restoring vision in individuals with optic atrophy.

In conclusion, artificial vision and prosthetics offer hope for individuals with optic atrophy by providing the possibility of restored vision. Visual prostheses, such as the Argus II system, have already shown promising results in improving limited vision. Ongoing research and advancements in technologies like optogenetics hold the potential for further enhancing the effectiveness of these treatments. While these solutions may not fully restore normal vision, they can significantly improve the quality of life for individuals affected by optic atrophy.

**Epidemiology**

The prevalence of optic atrophy varies widely. Optic atrophy was one of the five main causes of blindness in prevalence studies from Israel, Japan, Scotland, Zaire, and other countries.. In the Oman eye study, 5% of the blindness was attributable to this condition. In a study from Egypt, the age-adjusted blindness prevalence rates per 1000 persons showed that in an urban population, 4.1% of the blindness and in a rural population 1.2% of the blindness was attributable to optic atrophy.

In southern Germany, newly registered blindness-allowance recipients were analyzed for the causes of blindness. Among the 3531 individuals, the standardized incidence rate per 100,000 person-years for optic atrophy was 2.86 (SD 2.66-3.05). In a study from the Baltimore area of the United States, the prevalence of blindness due to optic atrophy in Whites was found to be zero, whereas it was 1.9% in African-Americans, giving a total prevalence of 0.8% across both groups. In another population-based study from the United States, 0.83% of individuals were found to be bilaterally blind. Of these, optic atrophy was responsible for bilateral blindness in 3 persons

**Differential diagnosis**

Certain conditions may mimic optic atrophy and have to be ruled out, including:

**Optic Nerve Pit**

These are congenital anomalies characterized by a greyish round or oval depression, usually on the temporal aspect of the optic nerve head. Serous macular detachments develop in nearly half of the cases.

**Myelinated Nerve Fibers**

Optic nerve myelination usually stops at the level of the lamina cribrosa. However, myelination extends into the retina following the nerve fiber bundles in certain individuals. Clinically they appear as feathery white patches that may obscure the disc margin and vessels. Visual field examination will show the enlargement of the blind spot.

**Optic Disc Drusen**

These are usually calcific deposits within the substance of the optic nerve head. They are bilateral in around 75% of the individuals. Usually asymptomatic, they may present with the occasional blurring of vision. In the pediatric age group, drusen are buried within the disc and appear as pseudopapilledema. In adult life, they get exposed, enlarge, and calcify. Later they may regress, leaving a pale disc. There is the filling of the optic cup, anomalous branching vasculature emanating from the central core, and hyaline bodies on the surface. Drusen also demonstrates fundus autofluorescence.

**Optic Nerve Hypoplasia**

This is a common optic nerve anomaly. The disc appears notably small due to the lack of axons passing through the optic nerve head.

**Brighter-Than-Normal Luminosity**

Using excessive illumination from the ophthalmoscope or slit lamp causes the disc to appear pale.

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**Ocular mucous membrane pemphigoid**

**Definition and description**

Mucous membrane pemphigoid (MMP) with ocular involve­ment, also known as ocular cicatricial pemphigoid, is a chronic autoimmune cicatrizing conjunctivitis. MMP can have serious, vision-threatening complications as well as systemic manifestations in the mouth, orophar­ynx, esophagus, skin, and genitoper­ineal region. Thus, it is important to diagnose MMP early in its clinical course, to treat it in a timely manner, and to prevent its adverse sequelae.

**Causes**

The cause of ocular cicatricial pemphigoid is an autoimmune type II hypersensitivity response.This autoimmune response occurs when a patient has a genetic predisposition and is exposed to an environmental trigger. Antibodies bind to surface antigens on the basement membrane of the conjunctiva. This causes recruitment and activation of complement proteins and inflammatory cytokines and leads to inflammation and scarring. There is evidence of an association to patients with HLA-DR4 and HLA-DQ7

**symptoms**

In people with ocular mucous membrane pemphigoid, both eyes are affected, becoming red at first. Later, the conjunctivae shrink, making it difficult to pull the upper or lower eyelid away from the eye. Much later, the eyes become dry.

The cornea (the clear layer in front of the iris and pupil) can become cloudy, preventing light from reaching the retina and decreasing vision.

The conjunctivae can scar and shrink, causing eyelashes to turn inward (see Trichiasis) and further damage the cornea.

**Diagnosis methods**

## 

| Category | Nonspecific Signs/Symptoms |
| --- | --- |
| General | Tearing, burning, light sensitivity, foreign body sensation |
| Conjunctival | Diffuse hyperemia, papillary reaction, keratoconjunctivitis sicca |
| Corneal | Punctate epithelial erosions, exposure keratitis, epithelial defects, peripheral infiltrates, ulcers, vascularization |
| Palpebral | Blepharitis, trichiasis, entropion, lagophthalmos |

## Table 1: Signs and Symptoms of MMP With Ocular InvolvementManagement

A multidisciplinary team that includes specialists in other medical fields such as rheumatology, hematology/oncol­ogy, and dermatology can be useful in managing MMP with ocular involve­ment, particularly its concurrent sys­temic symptoms. Such specialists can also contribute expertise in the use of systemic immunomodulatory agents, which are typically required to control MMP.

Topical therapy. Topical treatments such as artificial tears, punctal occlusion, steroids, tacrolimus, and cyclosporine A are useful adjuncts for relief of ocular surface disease symptoms. However, they have little effect in halting disease progression.

Systemic medications. Systemic anti-inflammatory and immunomod­ulatory medications are the mainstay of treatment for MMP with ocular involvement. A stepwise approach to systemic agents is taken depending on the severity of disease, presence of sight-threatening complications, and adverse effects of medications. The main categories of systemic agents are alkylating agents, antimetabolites, bio­logic immune modulators, interleukins, and T-cell inhibitors.

*In mild ocular disease,* dapsone is a common first-line anti-inflammatory and immunomodulatory agent. Its adverse effects, which include gastroin­testinal distress, anemia, hepatotoxicity, and leukopenia, can decrease adherence to treatment. Sulfapyridine and sulfasalazine are alternate first-line agents that have a lower adverse effects profile.

*In mild to moderate ocular disease,* mycophenolate mofetil is an effective and well-tolerated first-line antimetabolite agent for patients without sight-threatening complications. Methotrex­ate is another antimetabolite that has been recommended as a first-line agent because of its efficacy; however, it has more significant side effects, the most serious of which include hepatic and pulmonary fibrosis.

*In moderate to severe ocular disease,* **or in patients whose disease is** refractory to an adequate trial of first-line agents, systemic corticosteroids with concurrent immunosuppressives are commonly used. Cyclophospha­mide, an alkylating agent, typically in combination with corticosteroids, is a mainstay for patients who have severe disease with sight-threatening complications. However, it should be used judiciously because of its serious adverse effects, including pancytopenia and hepatotoxicity.

*In severe refractory ocular disease,* or in those with serious adverse reactions to prior levels of treatment, biologics such as etanercept, rituximab, and intravenous immunoglobulin are used.6 Finally, a recent study found repository corticotropin to be a well-tolerated alternate or adjunctive treatment in severe refractory MMP with ocular involvement.

Surgical management. It is important to note that surgery can be an inciting factor for further cycles of inflammation and cicatrization. There­fore, it is important to adequately con­trol active inflammation with medical therapy before proceeding with surgery.

Adjunctive surgical treatments for the sequelae of MMP with ocular involvement include eyelash epilation, entropion repair, tarsorrhaphy, and am niotic membrane graft. Procedures for more severe disease include limbal stem cell transplantation, mucous membrane autografts, and cultivated oral mucosal epithelial transplantation. The latter has been shown to be effective in treat­ment of ocular surface disease second­ary to limbal stem cell deficiency.

**In end-stage ocular disease, corneal transplants tend to fail due to graft vas cularization. However, some evidence supports the utility of keratoprosthesis, particularly the Boston keratoprosthesis type II, in select patients.**

## Table 2: Two Different Staging Systems for MMP With Ocular Involvement

| Stage | Foster System:  Clinical Signs | Mondino and Brown System:  Forniceal Depth Loss |
| --- | --- | --- |
| 1 | Subconjunctival scarring and fibrosis | <25% |
| 2 | Forniceal shortening | 25%-50% |
| 3 | Symblepharon formation | 50%-75% |
| 4 | Ankyloblepharon formation; corneal opacification | >75% |

## 

## **Prognosis**

Progression of ocular disease has been reported in 20% to 30% of patients undergoing systemic therapy. It is important to monitor progression with close interdisciplinary follow-up employing serial photos and detailed clinical documentation. Of note, a significant percentage of patients with MMP with ocular involvement demon­strate a clear and quiet conjunctiva despite clinical and histologic evidence of fibrotic progression. This makes management difficult, as injection cannot be used as a sign of disease activity. It has been proposed that monitoring inflammatory markers such as myeloperoxidase could be a quantitative way to track disease activity.

**The differential diagnosis for cicatriz­ing conjunctivitis includes the follow­ing conditions:**

* infections (e.g., adenovirus, *Chla­mydia trachomatis, Corynebacterium diphtheriae*)
* allergic eye disease (e.g., atopic kera­toconjunctivitis)
* drug-induced disorders (e.g., Stevens-Johnson syndrome, toxic epidermal necrolysis)
* autoimmune disorders (e.g., lichen planus, systemic lupus erythematosus, sarcoidosis scleroderma, linear IgA dermatosis, MMP with ocular involve­ment)
* chemical, thermal, or radiation injury
* medicamentosa
* pseudopemphigoid
* neoplasia
* ocular rosacea
* trauma

It is particularly difficult to distin­guish pseudopemphigoid from the ocular manifestations of MMP, as their presentations are identical. Pseudopem­phigoid is caused by long-term use of topical ophthalmic medications such as pilocarpine, timolol, and epineph­rine, and it resolves with cessation of the agent. Clinical information on laterality, skin and mucous membrane involvement, associated systemic dis­eases, medication use, and the presence of specific clinical signs can aid in dis­tinguishing the etiology of cicatrizing conjunctivitis.

Testing. In addition to clinical signs, a conjunctival biopsy sent to a pathol­ogy lab capable of performing direct immunofluorescence testing is used for definitive diagnosis. Conjunctival biopsy of an involved area, typically the inferior conjunctival fornix, is recommended, but biopsies of active oral lesions may also be taken. Linear staining of the epithelial basement membrane zone is consistent with diagnosis of MMP. A biopsy yielding negative immunohistochemical results has been reported in up to 60% of cases that were clinically consistent with the disease and its evolution. Negative biop­sy results can be attributed to inflam­matory destruction of the subepithelial membrane or therapies used prior to biopsy.

**Epidemiology**

Ocular cicatricial pemphigoid predominantly affects females (2:1 over males), and the age of onset is around 60 years of age or older. There is no racial predilection. Ocular cicatricial pemphigoid is considered a rare disease, and incidence is estimated to be about 1 per 10,000 to 50,000.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis includes all conditions that cause an asymmetric bilateral chronic conjunctivitis with conjunctival cicatrization. These include Stevens-Johnson syndrome, toxic epidermal necrolysis, trachoma, graft-versus-host disease, dry eye syndrome, history of adenoviral conjunctivitis, chemical burn, medicamentosa (from topical glaucoma medications and anti-viral medications for herpetic eye disease), atopic keratoconjunctivitis, radiation exposure, systemic lupus erythematosus, and Sjogren syndrome. A distinguishing clinical feature of OCP is a progressive symblepharon. Symblephara from the above etiologies may form and then remain stable. However, a few conditions in which progressive cicatrization occur include neoplasia, lichen planus, and paraneoplastic pemphigus

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**PAPILLEDEMA**

**Definition and description**

Papilledema refers to the swelling of both optic discs in your eyes due to increased intracranial pressure (intracranial hypertension). Cerebrospinal fluid (CSF) is constantly being released and then reabsorbed and normal levels help protect the brain from trauma.

The optic disc, or nerve head, is located where the optic nerve enters the back of the eye. The optic nerve is a pathway that connects the eye to the brain.

Intracranial hypertension usually results in bilateral papilledema (disc swelling of both optic discs). Swelling of the optic disc in only one eye (unilateral) is usually not the result of intracranial hypertension. Because of this, its name is optic disc edema.

Papilledema can be considered a medical emergency because intracranial hypertension can be serious, even potentially life-threatening.

Papilledema is more common in women. They’re usually 20 to 44 years old and are likely to be overweight (a BMI greater than 25) or obese (a BMI greater than 30). The incidence of papilledema in this group is 13 per 100,000.

The incidence is much lower among other groups in the U.S. The incidence of papilledema among the U.S. population as a whole is 0.9 per 100,000 people.

### **What causes papilledema?**

High intracranial pressure causes papilledema. Related issues may include:

* High blood pressure (malignant hypertension).
* Tumors.
* Infection, bleeding or inflammation in the brain or the meninges (the tissues that protect the brain and spinal cord).
* Cerebral venous sinus thrombosis. This condition refers to a blood clot in a vein in your brain.
* Iron-deficiency anemia.
* Medication usage. There have been links with retin-A and retinoids, toxic amounts of vitamin A, tetracycline antibiotics, COVID-19 and corticosteroids.
* Idiopathic intracranial hypertension (IIH). This condition literally means that the patient develops intracranial hypertension without a clear reason or identifiable cause. The word “idiopathic” means that there’s no clear reason or cause.

#### **Is papilledema hereditary?**

Papilledema isn’t hereditary.

#### **Do migraines cause papilledema?**

Migraines don’t cause papilledema.

**Signs and symptoms**

You may have no symptoms (be asymptomatic), though you may have:

* Headaches: Headaches related to papilledema may be worse in the mornings and when you’re lying down.
* Transient visual obscurations: These events are periods of about five to 15 seconds where your vision gets blurry, goes gray, or blacks out. An obscuration is similar to what happens during a total eclipse when the moon blocks the sun from your sight. These events usually happen when you change posture. You can have these events in both eyes (bilateral) or in only one eye (unilateral).
* Double vision (diplopia): This may happen if the intracranial hypertension results in a cranial nerve palsy that impairs the eye muscles.
* Nausea and vomiting.
* Neurological symptoms: These may include problems with movement or thinking.

The vision loss worsens as the condition progresses.

**Diagnosis methods**

You may find out you have papilledema while your provider is evaluating you for other conditions, like illnesses that cause headaches and vision problems.

Your eye doctor will perform an eye exam with additional tests that may include visual field testing. They’ll be able to see if your optic disc looks swollen.

If your optic disc seems to be swollen, your provider will send you for an imaging examination, most likely a magnetic resonance imaging (MRI) scan.

Your provider may order a lumbar puncture (spinal tap).

Other tests may include blood tests to determine levels of iron and other substances in your blood.

### **How is papilledema graded (staged)?**

Providers might use a version of the Frisén scale to grade or stage papilledema (that rates severity). The stages run from Grade 0 where the disc is normal but the edges may be blurred to Grade V where the whole disc is elevated, and you can’t see the blood vessels in the disc or those leaving the disc.

## **Management and Treatment**

For papilledema due to idiopathic intracranial hypertension, your provider may prescribe a carbonic anhydrase inhibitor such as acetazolamide. They may also work with you to achieve and maintain a healthy weight as a long-term strategy.

If these methods don’t work, your provider may suggest surgical procedures to relieve and decrease intracranial hypertension.

For papilledema due to other causes, your provider will treat the underlying cause.

It’s important to treat papilledema because untreated papilledema can lead to partial or complete blindness in one or both eyes.

## **Prevention**

You can reduce your risk of developing papilledema by:

* Keeping your blood pressure under control (avoid malignant hypertension).
* Achieving and keeping a healthy weight.

Regular eye exams are important. As with most medical conditions, early diagnosis leads to better outcomes.

## **Outlook / Prognosis**

If you and your provider catch papilledema early, the outlook is good. Your provider will treat the condition primarily by addressing the issue that caused it.

It’s important to treat papilledema because of the potential for blindness and other neurologic effects.

## **Living With**

### **When should I see my healthcare provider about papilledema or its potential causes?**

If you have new or worsening headaches, along with nausea and vomiting, you should seek immediate medical care. You should have any change in your vision assessed by a healthcare provider.

## **Differential Diagnoses**

* Acute Complications of Sarcoidosis
* Adult Optic Neuritis
* Anterior Ischemic Optic Neuropathy (AION)
* Central Retinal Vein Occlusion (CRVO)
* Compressive Optic Neuropathy
* Hypertension
* Idiopathic Intracranial Hypertension (IIH)
* Optic Disc Drusen (Pseudopapilledema)  
  Causes of pseudopapilledema include optic disc drusen and myelinated nerve fibers.
* Scleritis in Emergency Medicine
* Thyroid Ophthalmopathy
* Toxic/Nutritional Optic Neuropathy
* Toxoplasmosis
* Uveitis Classification
* Vogt-Koyanagi-Harada (VKH) Disease

**EPIDEMIOLOGY**

* Papilledema from various causes of IH may develop at any age, in either sex, and in any racial or ethnic group. Although high ICP can occur in infants and very young children, open fontanels may mitigate the development of papilledema in these patients despite IH.
* In contrast, idiopathic intracranial hypertension (IIH) predominantly affects obese women of childbearing age . In the USA, the annual incidence per 100,000 persons has been estimated to be 0.9 in the general population and 3.5 in females 15–44 years of age Durcan et al estimated that the incidence of IIH is up to 13 per 100,000 in obese women aged 20–44 years of age who were ≥10% overweight and up to 19.3 per 100,000 if ≥20% over ideal weight. The incidence of IIH varies from country to country, probably related to the prevalence of obesity. The incidence was 1.56/100,000 persons/year, 2.86/100,000 in women, and 11.9/100,000 in obese women in the Sheffield UK study.The reported annual incidence of IIH in Mid

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**PROPTOSIS**

**Definition and description**

Proptosis is when one or both of your eyes bulge from their natural position. The condition can affect your appearance, leaving you with a startled expression that doesn’t go away.

Proptosis can also make it difficult for you to blink. When you can’t blink, the protective outer layer of your eyes (corneas) doesn’t receive the lubrication it needs to do its job. You may be at risk for cornea damage. Some people experience other complications, like low vision or double vision (diplopia).

Other names for proptosis include bulging eyes, protruding eyes and exophthalmos.

## **Causes of Proptosis**

**Understanding the underlying causes of proptosis is essential for effective treatment. The causes can be broadly categorized into systemic diseases, local orbital diseases, and other miscellaneous conditions.**

## **Symptoms**

### **What are the signs of proptosis?**

When your eyes bulge, the white part of your eyes is more visible. In addition, your eyeballs push forward from your eye sockets. You may be able to see less of your upper eyelids because your eyes look like they’re wide open.

Other symptoms of a bulging eye that you may experience include:

* A gritty sensation when you move your eye.
* Dry, irritated or watery eye (epiphora).
* Eyelid swelling or redness.
* Muscle tightness that may prevent you from moving your eye.
* Sensitivity to light.
* Headache.
* Fever.

### Are there symptoms I should be concerned about?

Symptoms of proptosis that need a prompt evaluation from a healthcare provider include:

* A throbbing sensation in your eyes.
* Eye bulging that comes on suddenly.
* Pain or redness.
* Symptoms in one eye.
* Loss or decrease of vision.
* Blurred or double vision.
* Difficulty focusing on objects.

### What causes bulging eyes?

The most common cause of bulging eyes is an autoimmune disease in which your body’s immune system attacks cells in your thyroid gland and the tissue behind your eye. Proptosis in people with thyroid issues is also called thyroid eye disease (TED). People who have thyroid issues are the most likely to experience bulging eyes. Proptosis causes include:

* Graves’ disease.
* Hyperthyroidism (overactive thyroid gland).
* Eye socket infections.
* Injuries, especially if they cause bleeding behind your eye.
* Tumors, which may include neuroblastoma and some soft tissue sarcomas.

### **Complications and long-term effects of proptosis**

Most people don’t experience complications or long-term effects. In rare cases, double vision or vision loss may be permanent. It’s also possible to experience cornea damage if you can’t blink and lubricating eye drops aren’t effective.

## **Diagnosis and Tests**

Your healthcare provider will ask about your symptoms and medical history to determine the potential causes of your bulging eyes.

They’ll also perform an eye exam that may include:

* Using a slit lamp to magnify your eye’s surface and structures.
* Assessing eye and eyelid movement.
* Checking for redness, soreness and irritation.

#### What other tests might I need?

Your healthcare provider may perform or recommend other tests, including:

* Exophthalmometry: This test uses a special instrument to measure how far your eyeball protrudes from your eye socket.
* Blood tests: This will likely include a workup for thyroid disease.
* Imaging studies: Imaging tests, like an MRI (magnetic resonance imaging) or a CT scan (computed tomography scan), can check for bleeding, tumors or signs of infection.
* Other lab tests: Lab tests, like a blood or tissue culture, can confirm or rule out an infection.

## **Management and Treatment**

Your treatment may include:

* Artificial tears, including drops or gel, to relieve dry eyes and protect your corneas.
* Antibiotics if you have an infection.
* Medical treatments for underlying conditions, like medications for hyperthyroidism.
* Intravenous (IV) medications like corticosteroids (anti-inflammatory medication) or teprotumumab (Tepezza®) for thyroid eye disease.

#### Are there other nonsurgical treatments for bulging eyes?

Other therapies may include:

* Double vision treatments: These treatments include prisms that attach to your glasses and redirect light as it enters your eyes.
* Immunosuppressive drugs: These drugs may lessen the impact of immune system attacks on your eyes.
* Corticosteroids: You may receive steroids by injection or through a vein in your arm to relieve swelling or restore eyesight.

#### Will I need surgery?

You may need surgery to:

* Create more space behind your eye in the eye socket.
* Treat double vision.
* Protect your corneas if you can’t fully close your eyelids.
* Remove a tumor.

## **Outlook / Prognosis**

### What is the outlook for people with protruding eyes?

You’re more likely to have good outcomes if you receive timely treatment to address the cause of your symptoms. Getting the right therapies for your needs can also help you avoid complications.

Yes. But even with successful treatment, it may take a while for your eye to return to its usual position. In some cases, it takes years.

## **Prevention**

Steps you can take to prevent proptosis from getting worse include:

* Keeping thyroid levels in check: If you have thyroid disease, follow your healthcare provider’s care instructions. Take daily medications and stay current with blood testing to check thyroid levels.
* Quitting smoking: Smoking can make proptosis treatments less effective. Quitting can have a significant and positive impact on your body’s response.

## **Living With**

Protruding eyes can change your appearance and cause issues with your confidence and self-esteem. If it affects your vision, you may experience unexpected changes in your daily life. These changes can leave you feeling upset, anxious or depressed.

#### How can I cope with challenges?

Support from a mental health professional or social worker can help you feel better:

* Mental health professionals may use cognitive behavior therapy (CBT) to help you find methods for coping with negative thoughts and feelings.
* Social workers can connect you with local resources in your area, including support groups, or virtual resources if you’re unable to drive due to vision issues.

## **Epidemiology**

### Frequency

Five percent of the general population is affected by thyroid autoimmunity problems.

United States

Bartley et al reported a frequency of 2.9 cases per 100,000 population per year in men and 16 cases per 100,000 population per year in women. They also observed a bimodal distribution in both sexes, with women showing one peak at age 40-44 years and the other peak at age 60-64 years. In men, the bimodal occurrence was at age 45-49 years and age 65-69 years. Both peaks incidences in men were 5 years later than in women.

International

In a nationwide registry-based cohort study conducted in Denmark between 2000 and 2018, researchers investigated the incidence of thyroid eye disease (TED), strabismus, and surgical interventions related to TED. The study included a cohort of approximately 4.3 million individuals aged 18 to 100 years with no prior TED diagnosis each year. The total observation time amounted to 8.22 million person-years (women accounting for 4.18 million person-years and men for 4.04 million person-years).

Over 19 years, there were 4106 new cases of thyroid eye disease (TED) documented, with a prevalence of 81.4% among women and 18.6% among men. The average annual incidence rate of TED was 5.0 per 100,000 person-years, significantly higher in women (8.0 per 100,000 person-years) than in men (1.9 per 100,000 person-years), resulting in a notable 4:1 ratio. The average age of onset for TED was 51.3 years. At the time of diagnosis, 14.9% of patients were euthyroid, 11.6% were hypothyroid, and 73.5% were hyperthyroid. In patients who were euthyroid, the 4-year cumulative incidence for antithyroid medication was 41%, whereas for L-thyroxine, it was 13%. The prevalence of strabismus in TED patients within 4 years was 10%. Surgical interventions, including strabismus surgery and orbital decompression, had a 4-year cumulative rate of 8% and 5%, respectively. Men had a higher likelihood of undergoing strabismus surgery compared to women 4 years post-TED diagnosis.

## **Differential Diagnoses**

* Ptosis (Blepharoptosis) in Adults
* Anophthalmos
* Apex Orbital Fracture
* Carotid-Cavernous Fistula (CCF)
* Orbital Cavernous Hemangioma
* Congenital Ptosis (Drooping Eyelid)
* Dacryoadenitis
* Duane Syndrome
* Orbital Floor Fractures (Blowout Fractures)
* Globe Retraction
* Horner Syndrome
* Juvenile Glaucoma
* Ophthalmologic Manifestations of Leukemias
* Medial Wall Orbital Fracture

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### 

**PROSTATITIS**

**Definition and description**

Prostatitis is a condition of the prostate gland most often linked with swelling and irritation, called inflammation. Prostatitis can make it painful or hard to urinate. It also may cause pain in the groin, pelvic area or genitals. Bacterial infections cause some but not all prostatitis.

The prostate gland, about the size of a walnut, sits just below the bladder in people assigned male at birth. It surrounds the top part of the tube that drains urine from the bladder, called the urethra. The prostate and other sex glands make the fluid that carries sperm during ejaculation. This fluid is called semen.

### **Types**

There are four main types of prostatitis:

* **Acute bacterial prostatitis.** Bacteria causes this infection of the prostate. It most often has sudden, serious symptoms.
* **Chronic bacterial prostatitis.** This is a bacterial infection that lasts or comes back. The symptoms often are less serious than are those of acute bacterial prostatitis.
* **Chronic prostatitis, also called chronic pelvic pain syndrome.** This causes pelvic pain and urinary tract symptoms that last or come back. But there's no sign of infection.
* **Asymptomatic inflammatory prostatitis.** This has signs of an inflamed prostate with no urinary symptoms.

## **Causes**

**Prostate gland**

Causes depend on the type of prostatitis.

* **Acute bacterial prostatitis.** Common strains of bacteria are most often the cause. The infection may have spread from other parts of the urinary or reproductive systems.
* **Chronic bacterial prostatitis.** This most often has the same cause as acute bacterial infection. It may happen when treatment for an acute infection isn't long enough or fails to kill all the bacteria.
* **Chronic prostatitis, also called chronic pelvic pain syndrome.** Research suggests that the cause may involve several factors. These include an earlier infection, a condition of the nervous system or immune system, psychological stress, or issues with hormones.
* **Asymptomatic inflammatory prostatitis.** This has no known cause. It may show up during an exam for other medical conditions.

## **Risk factors**

Risk factors for prostatitis include:

* Young or middle adulthood.
* Earlier prostatitis.
* Infection of the urinary or reproductive system.
* HIV infection or AIDS.
* Use of a tube put into the urethra to drain the bladder, called a urinary catheter.
* Having a sample of prostate tissue taken for study in a lab, called a biopsy.

Other risk factors for chronic prostatitis, also called chronic pelvic pain syndrome, may include:

* Mental stress.
* Nerve damage in the pelvic region due to surgery or injury.

**Signs and symptoms**

Symptoms of prostatitis depend on the type of condition. They may include:

* Pain or burning feeling when urinating, called dysuria.
* Trouble urinating, such as dribbling or trouble starting a stream of urine or keeping it going.
* Urinating often, mostly at night, called nocturia.
* Urgent need to urinate.
* Cloudy urine.
* Blood in the urine.
* Pain in the belly, groin or lower back.
* Pain in the area between the scrotum and rectum, called the perineum.
* Pain or discomfort of the penis or testicles.
* Painful ejaculation.
* Fever, chills, muscle aches and other flu-like symptoms with acute bacterial prostatitis.

## **Diagnosis**

Several conditions can cause symptoms like those of prostatitis. Your healthcare professional may send you to a specialist in urinary and reproductive system conditions, called a urologist. Diagnosis involves a physical exam, review of your symptoms and medical history, and tests.

### **Test for diagnosing bacterial infections**

Diagnostic tests to assess for infection may include:

* **Digital rectal exam.** With this procedure, a healthcare professional puts a gloved finger that's oiled into your rectum to see if there's inflammation of the prostate.
* **Urine test.** A urine sample is tested for infection and what type it is.
* **Blood test.** Blood samples may show signs of infection and other prostate conditions.
* **Prostatic specimen test.** Sometimes, a healthcare professional gently massages the prostate during a rectal exam. This is to release prostate fluid into the urethra. A urine sample collected after the massage is tested to check for infection in the prostate fluid. This test is not for acute bacterial prostatitis because it can spread germs in the blood.

### **Other tests**

If the first tests show no sign of infection, you may have other tests, including:

* **Urodynamic tests.** These tests measure how well the bladder and urethra hold and release urine. These tests can help show the source of issues with urinating.
* **Imaging.** Imaging tests can show something that isn't usual in the prostate and growths or other issues in the pelvic region that may be causing pain.

## **Treatment**

Treatment for prostatitis depends on the type you have and your symptoms.

### **Treating infection**

For acute or chronic bacterial prostatitis, you take antibiotics. Acute prostatitis may need antibiotics given through a tube in a vein, called an IV, in the hospital for a short time.

The course of antibiotic treatment is most often 4 to 6 weeks. Sometimes it can be longer. Take all the medicine to get rid of the infection and lower the risk of chronic bacterial prostatitis.

### **Treating urinary symptoms**

Medicines called alpha-blockers help relax the bladder neck and the muscle fibers where the prostate joins the bladder. This treatment might ease symptoms, such as pain while urinating or trouble urinating.

Alpha-blockers most often treat people with chronic prostatitis, also called chronic pelvic pain syndrome. Alpha-blockers also can ease urinary symptoms of bacterial infections.

### **Treating pain**

Your healthcare professional may prescribe pain medicine or suggest medicines you can get without a prescription. These include acetaminophen (Tylenol, others) and ibuprofen (Advil, Motrin IB, others).

### **Managing psychological symptoms**

Your healthcare professional may advise that you see a mental healthcare professional. This can help you manage stress, depression or worry that may be linked with long-term pain.

**Lifestyle and home remedies**

Do the following to help ease some symptoms of prostatitis:

* Soak in a warm bath, called a sitz bath, or use a heating pad.
* Don't have or limit alcohol, caffeine, and spicy or high-acid foods. They can irritate the bladder.
* Drink plenty of water. This will make you urinate more and help flush bacteria from your bladder.

## **Alternative medicine**

Alternative therapies that show some promise for easing symptoms of prostatitis include:

* **Biofeedback.** You use this mind-body technique to help you control some of your body's work, such as heart rate and muscle responses. During biofeedback, you're connected to electrical pads that help you learn how your body responds.
* **Acupuncture.** This treatment involves putting very thin needles through your skin at certain points on your body. Acupuncture may help ease pain.
* **Herbal treatments.** Some studies suggest that rye grass pollen extract, also called cernilton, may help manage the pain of chronic prostatitis, also called chronic pelvic pain syndrome. There isn't enough proof that other herbal treatments ease symptoms of prostatitis.

Talk about your use of alternative medicine practices and herbal treatments with your healthcare professional before trying any.

**Complications**

Complications of acute or chronic prostatitis can include:

* Bacterial infection of the blood, called bacteremia.
* Irritation of the coiled tube attached to the back of the testicle, called epididymitis.
* Pus-filled cavity in the prostate, called a prostatic abscess.
* Infection that spreads to the upper pelvic bone or lower spine.

Complications of chronic prostatitis/chronic pelvic pain syndrome may include:

* Worry or depression.
* Sexual dysfunction, such as not being able get and keep an erection, called erectile dysfunction.
* Changes in sperm and semen that may affect having children, called infertility.

There's no proof that prostatitis can lead to prostate cancer. Researchers are looking into whether long-term irritation of the prostate is a risk factor for cancer.

### **When to see a doctor**

Several conditions can cause symptoms like those of prostatitis. Get a diagnosis and treatment as soon as possible.

Get care right away if you:

* Are not able to pass urine.
* Have a fever and trouble urinating or pain while urinating.
* Have blood in your urine.
* Have a lot of discomfort or pain in the pelvic area or genitals.

**Differential diagnosis**

In addition to prostatitis, other conditions to consider include the following:

* Benign prostatic hyperplasia
* Chronic pain syndromes (ie, inflammatory bowel disease)
* Cystitis
* Erectile dysfunction
* Prostatic Abscess
* Pyelonephritis
* Prostate cancer
* Radiculopathies
* Testicular cancer
* Urolithiasis

## Differential Diagnoses

* Anal Fistulas and Fissures
* Hemorrhagic Cystitis, Noninfectious
* Inflammatory Bowel Disease
* Male Urethritis
* Mechanical Back Pain
* Rectal Foreign Bodies
* Urinary Incontinence
* Urinary Tract Obstruction Management in the ED
* Urinary Tract Infection (UTI) in Males

## **Epidemiology**

### United States statistics

Prostatitis is one of the most common diseases seen in urology practices in the United States, accounting for nearly 2 million outpatient visits per year.The diagnosis is made in approximately 25% of male patients presenting with genitourinary symptoms.

Approximately 8.2% of men have prostatitis at some point in their lives.Among the 4 categories of prostatitis, the most common is chronic prostatitis/chronic pelvic pain syndrome, accounting for 90-95% of prostatitis cases. Acute bacterial prostatitis and chronic bacterial prostatitis each make up another 2-5% of cases. Because asymptomatic inflammatory prostatitis is only identified incidentally, its overall prevalence is difficult to estimate.

### International statistics

The prevalence of prostatitis amongst men in North America, Europe, and Asia is estimated to be between 2-10%.

### Age-related demographics

In patients younger than 35 years, the most common variant of the syndrome is acute bacterial prostatitis. HIV-related disease is also predominantly seen in younger patients.

Among older patients, nonbacterial prostatitis (National Institutes of Health [NIH] types II and IV) are the most common. Of importance, rare causes of prostatitis should be sought during evaluation. According to case reports of Wegener granulomatosis in the fourth and fifth decades of life, prostatitis can be a presenting feature of Wegener granulomatosis and a clinical manifestation of relapse.

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**Pterygium**

**Definition and description**

A pterygium (*ter-IJ-ee-um*) is a raised, fleshy growth on your eye’s conjunctiva, the clear membrane that covers the white part of your eye. The growth may look whiteish or pinkish and may have visible blood vessels in it. It typically starts in the corner of your eye and grows toward your iris (the colored part).

Pterygium typically has a triangular or wing-like shape. The word, “pterygium,” comes from the Greek words *pteryx*, meaning “wing,” and *pterygion*, meaning “fin.” Another name for pterygium is surfer’s eye. This is because surfers are often in the elements that cause pterygium — sun, wind, sand and salt.

#### Who gets pterygium (surfer’s eye disease)?

Surfers do get pterygia (the plural of pterygium) more often. But anyone can get one if they spend a lot of time outdoors without eye protection. It’s more common in older adults, who’ve had more exposure to the elements over time, and also in those who live near the equator, where sunlight is more intense.

A pterygium isn’t an invasive growth. It’s not cancerous, and it won’t spread to other nearby tissues. But it can continue to grow across the surface of your eye. If it reaches your cornea (the clear part that covers the colored part), it can damage your vision. In this case, you might need surgery to remove it.

### **What causes a pterygium eye?**

Pterygium is an overgrowth of your conjunctiva tissue. Causes include:

* Long-term exposure to the sun’s ultraviolet (UV) light.
* Chronic irritation from hot and dry weather, wind and dust.

Other possible risk factors include:

* Age over 60.
* Genetics.
* Vitamin A deficiency.
* Human papilloma virus (HPV).

**Risk factors**

The primary risk is recurrence (return of the pterygium after surgery). If this happens, you may need another surgery. Fortunately, with new and improved surgical techniques, rates of pterygium recurrence are declining. Current research suggests the rate of recurrence is somewhere between 2% and 15%.

Recurrence is more likely to occur if you don’t take your prescribed eyedrops as directed, or if your eye has direct sun exposure after surgery. It’s important to follow your provider’s instructions on this. You can still spend time outside, but if you do, make sure to wear protection, like a hat or sunglasses.

Some of the medications that providers prescribe to prevent recurrence can also have side effects. Antimetabolites like mitomycin can cause “melting” or thinning of your sclera (scleromalacia) after pterygium surgery. Steroid eye drops can raise your eye pressure or lead to cataracts forming.

### **What symptoms can a pterygium cause?**

You might not feel your pterygium at first. You might only notice a raised, fleshy, wedge-shaped growth at the corner of your eye. When symptoms do develop, they can be mild to severe. They might include:

* Inflammation and swelling (conjunctivitis).
* Redness or bloodshot eye.
* Dry eye.
* Watery eye.
* A feeling like there’s something in your eye.
* Eye itching.
* Eye burning.
* Eye pain.

Over time, you might also notice:

* An increase in the size and spread of the lesion.
* Difficulties or discomfort with contact lenses.
* Vision changes, like blurred or double vision.

Not everyone develops these symptoms. Some pterygia grow more than others.

**Diagnosis methods**

Your eye care provider can diagnose a pterygium with a slit lamp exam. A slit lamp is a type of microscope that focuses a narrow line (slit) of bright light on your eye. It helps your provider look at the front and inside of your eye. This is part of a standard eye exam. They’ll recognize a pterygium on sight.

## **Management and Treatment**

Pterygium treatment depends on how much it’s affecting you. If it’s not causing you any symptoms, it probably doesn’t need treatment. But your eye care provider will keep an eye on it. They’ll schedule regular eye exams to check on how the pterygium is growing and whether it’s affecting your vision.

If the pterygium irritates your eye, your provider might prescribe:

* Over-the-counter (OTC) eye drops or eye ointments.
* A short course of steroid eye drops for severe symptoms.

Providers also recommend wearing hats and wraparound UV sunglasses to protect your eyes from further UV damage. This may help slow the growth of your pterygium or prevent it from growing faster. If your pterygium is growing toward your cornea, they’ll recommend removing it before it gets there.

#### How do you get rid of a pterygium?

The only way to get rid of a pterygium is with eye surgery. A pterygium won’t go away on its own. You may want to have your pterygium removed if it seriously irritates your eye, if it’s growing aggressively or if it’s already affecting your vision. You may also choose to have it removed for cosmetic reasons.

### What’s involved in pterygium surgery?

An ophthalmologist can remove your pterygium. If you don’t have one, your optometrist can refer you.

During pterygium surgery, your ophthalmologist will:

* Give you anesthesia, so you won’t feel anything.
* Remove the abnormal tissue from the surface of your eye.
* Cover the hole left in your conjunctiva with other tissue.

Surgeons use different methods for patching the hole left after they remove your pterygium.

Options include:

* Conjunctival autograft. Your surgeon may take a piece of conjunctiva tissue from elsewhere on your eyeball, usually from behind your upper eyelid, to patch the hole. This method leaves another wound behind your eyelid, but it’s well protected there and heals on its own.
* Amniotic tissue graft. Surgeons use donated amniotic membrane tissue (from a placenta) to cover the hole while it heals. This tissue acts as a bandage that protects your conjunctiva until it can grow back to cover the hole. This method is helpful when the gap is too big for an autograft.

Pterygium surgery takes about an hour. After surgery, you’ll wear an eye patch for a few days while you recover. It takes four to six weeks for your eye to heal completely.

Your provider will prescribe medications to take home with you, including antibiotics to prevent infection and steroid eye drops to prevent the pterygium from growing back.

## **Prevention**

You can lower your risk of developing a pterygium — and slow its growth if you have one — by protecting your eyes from the elements, especially sunlight. Wear UV protection sunglasses when you’re in the sun. Protect your eyes from dry, windy, dusty climates by lubricating them with artificial tears.

## **Outlook / Prognosis**

A pterygium won’t go away unless a surgeon removes it. It’s likely to grow slowly throughout your life. Most people won’t need treatment for a pterygium. If it irritates your eye, you can usually manage these symptoms with over-the-counter medications. If it’s more than a minor nuisance, you can remove it.

There’s a small chance your pterygium will come back after surgery. If it does, it usually starts to appear within four months, and no later than 12 months after surgery. If you need to have another surgery, your surgeon will take extra measures to make sure the pterygium doesn’t come back again.

## **Living With**

### When should I see my healthcare provider about my pterygium?

Your provider will want to see you regularly to check on your pterygium and measure its growth — usually about once a year, unless it grows faster or slower than normal. Make sure to see them sooner if you start to develop new symptoms, or if your current medications aren’t working for you anymore.

While evaluating a case of pterygium, the following conditions should be ruled out:

Pseudopterygium

It is a fold of bulbar conjunctiva which is attached to the cornea. It is an inflammatory reaction where adhesions are formed between bulbar conjunctiva and cornea. A 'probe' test can be performed where the probe can be passed beneath the tissue in pseudopterygium but not in pterygium. It can appear in any quadrant of the eye and is non-progressive in nature.

Pinguecula

It is a yellowish colored growth on the bulbar conjunctiva close to the limbus. It is said to be a deposit of fat, protein or calcium. It is commonly found nasally. The patient is usually asymptomatic but may present with mild redness and foreign body sensation when inflamed.

Nodular Episcleritis

A discrete area of inflammation in episcleral tissue is seen, which is elevated. Only vascular congestion may be present in cases of simple episcleritis. If phenylephrine 2.5% is instilled, there will be blanching of the superficial episcleral vascular network. Usually, this condition is mildly painful, unlike pterygium.

Marginal Keratitis

It is an inflammatory disease of the peripheral cornea. Stromal infiltrates with the breakdown of the epithelium, and ulceration is seen. Infiltrate on the corneal surface is separated from limbus by a clear zone.

Phlycten

It is a result of delayed-type immune reaction leading to nodular inflammation of the conjunctiva.

Conjunctival carcinoma in situ/ Bowen epithelioma

It is a rare presentation. It presents as a gelatinous-appearing mass.

Ocular surface squamous neoplasia (OSSN)

**Possible complications**

### What happens if a pterygium isn’t removed?

Most pterygia will continue to grow, but some grow much more slowly than others. It’s possible that your pterygium will never bother you much. Because of the risk that it might grow back, healthcare providers don’t recommend surgery for a pterygium that’s small and doesn’t cause symptoms.

However, a pterygium that keeps spreading across your eye can cause problems. It may irritate your eye or obscure your vision. If it reaches your cornea, it can pull at it and change the shape, causing astigmatism. It can also scar your cornea, which can affect your vision even after it’s removed.

**Epidemiology**

Pterygium is a degenerative disorder of the conjunctiva. It is usually seen as a triangular fleshy fibrovascular proliferation from the bulbar conjunctiva onto the cornea, located mostly on the nasal side. Though it occurs worldwide, its prevalence is high in the “pterygium belt” between 30 degrees north and 30 degrees south of the equator.[1](https://pmc.ncbi.nlm.nih.gov/articles/PMC5968422/#B1) The prevalence of pterygium is reported to be 3% in Australians, 23% in blacks in United States, 15% in Tibetans in China, 18% in Mongolians in China, 30% in Japanese and 7% in Singaporean Chinese and Indians.

In a population-based study from rural central India, prevalence of pterygium increased from 6.7±0.8% in the age group from 30–39 years to 25.3±2.1% in the age group of 70–79 years. Three population based studies have described the incidence of pterygium. Barbados eye study has described the nine year incidence of pterygium to be 11.6% (95% CI, 10.1–13.1), the Beijing Eye Study described the 10 year incidence of pterygium in the adult Chinese population to be 4.9%, and the five year cumulative incidence in Bai Chinese population in a rural community was 6.8% (95% CI, 5.2–8.4)

**DIFFERENTIAL DIAGNOSIS**

differential diagnosis of pterygium includes several ocular surface conditions that can mimic its appearance or share similar clinical features. Key differential diagnoses are:

* Pinguecula: A common, benign, yellowish, elevated conjunctival lesion caused by actinic damage, typically confined to the perilimbal conjunctiva and not extending onto the cornea, unlike pterygium.
* Pseudopterygium: An inflammatory adhesion of the conjunctiva to the cornea, which can occur anywhere around the corneal limbus, unlike pterygium that is characteristically located nasally or temporally at the 3 and 9 o’clock positions.
* Ocular Surface Squamous Neoplasia (OSSN): Includes conjunctival intraepithelial neoplasia and squamous cell carcinoma; these malignant or premalignant lesions may resemble pterygium but often show leukoplakia, pigmentation, irregular feeder vessels, atypical elevation, or rapid growth. High-resolution anterior segment OCT and histopathology help differentiate OSSN from pterygium
* Limbal Dermoid: A congenital benign tumor presenting as a white or yellowish mass at the limbus, often with hair follicles, which can be confused with pterygium in atypical presentations.
* Corneal Phlyctenule: Small, raised, inflammatory nodules on the cornea or limbus that can mimic early pterygium.
* Pannus: Superficial neovascularization and fibrosis of the cornea, often secondary to chronic inflammation, may resemble a pterygium but lacks the typical fibrovascular growth pattern.
* Stevens-Johnson Syndrome and Symblepharon: Chronic conjunctival scarring and adhesions can produce conjunctival changes that may mimic or complicate pterygium diagnosis.
* Fuchs’ Superficial Marginal Keratitis and Terrien’s Marginal Degeneration: Corneal inflammatory and degenerative conditions that may present with peripheral corneal changes resembling pterygium

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**PTOSIS**

**Definition and description**

Ptosis is a condition in which your upper eyelid droops, sags or falls over your eye. It usually happens because your levator muscle — the muscle that lifts your eyelid — doesn’t work as it should. The condition can limit your vision or block it completely, depending on how much your lid droops. Ptosis of the eyelid can affect one or both upper eyelids. It can also be worse in one eye than the other.

Other names for the condition include blepharoptosis, upper eyelid ptosis or droopy eyelid.

#### **Types of ptosis**

Ptosis can affect both children and adults.

* Congenital ptosis: Congenital ptosis means your child was born with the condition. Problems with the development of the levator muscle cause congenital ptosis.
* Acquired ptosis: This type of ptosis affects adults later in life. It occurs when your levator muscle weakens or separates away from your eyelid.

### **What causes ptosis?**

Ptosis causes depend on the type. Some babies are born with ptosis in one or both eyelids (congenital ptosis).

Ptosis can occur later in life (acquired ptosis) if the muscles or ligaments that normally raise your eyelid are weakened by injury or disease. Sometimes, the drooping is a result of damage to the nerves that control your eyelid muscles.

Diseases and conditions that may result in ptosis include:

* Stye.
* Horner syndrome.
* Myasthenia gravis.
* Stroke.
* Tumor.
* External ophthalmoplegia.

Most ptosis just happens with aging. As you age, the skin and muscles of your eyelids stretch and weaken. Sometimes, previous eye surgery speeds up this change because the instruments used to keep your eye open during surgery can stretch your eyelid.

### **What happens if ptosis is left untreated?**

If your child has congenital ptosis, the sooner you have it treated, the better. If left untreated, it could impact their vision development and lead to other issues. Mild acquired ptosis is less likely to cause vision problems, and you may not need to seek treatment. But severe ptosis can cause serious complications if you don’t treat it. Complications of ptosis may include:

* Astigmatism: When your eyelid puts pressure on the front of your eye, it can change the shape of your eye. This can cause distortion in your vision (your vision may be stretched or wavy).
* Amblyopia: Astigmatism and other refractive errors (issues focusing due to a need for glasses) can cause amblyopia, or lazy eye.
* Chin-up position: When your child has to tilt their chin up to be able to see beyond their drooping eyelids, it can cause neck problems, tighten forehead muscles and developmental delays.

### **What are the symptoms of ptosis?**

You can typically tell if you or your child has ptosis by your eyelid’s appearance. It may cover only your upper eye, or it may cover your entire pupil. Other ptosis symptoms may include:

* Excessive rubbing of your eyes.
* Increased tearing.
* Decreased or impaired vision.
* Tiredness and achiness around your eyes.
* Children may tip their heads back to see.

## **Diagnosis and Tests**

Healthcare providers can typically detect ptosis by the appearance of a drooping eyelid. If both eyelids are affected, it may be more difficult to diagnose. Your healthcare provider will perform a physical exam. They may refer you to an eye care specialist (ophthalmologist) who will perform an eye exam and may request additional tests.

#### **What tests will be done to diagnose this condition?**

Tests that your eye care provider may perform include:

* Slit lamp examination.
* Visual field testing.
* Ocular motility (eye movement) test.
* Tensilon test (uses the drug Tensilon, also known as edrophonium, to diagnose myasthenia gravis).

## **Management and Treatment**

Ptosis treatment usually depends on how well your eyelid muscles are functioning. If the condition doesn’t affect your vision and the appearance doesn’t bother you, you might not need treatment at all.

If ptosis causes a problem with vision, appearance or both, your eye care specialist may recommend treatment. The type of treatment depends on whether the ptosis is caused by a disease or by aging. Treating ptosis caused by aging usually involves surgery.

#### **Ptosis surgery**

Ptosis surgery is performed under local anesthesia with sedation (you’re awake but you don’t feel the procedure). The types of surgery to repair the droopy lid include the following:

* Your surgeon makes an opening in the skin of your upper eyelid. This allows the surgeon to find the small muscle that raises your eyelid. The surgeon places stitches to tighten this muscle and raise your eyelid. The incision in the skin of your eyelid is then closed with more stitches.
* The surgeon can perform the entire surgery from underneath your eyelid. In this case, your surgeon flips your eyelid and tightens the muscle from underneath. No skin incision is required for this approach.

After surgery, your surgeon will explain how to take care of your eye. It’s important to come back to your provider after surgery so they can check your results. Appointments are usually scheduled for several days to one week after surgery.

**Prevention tips**

### **Can ptosis be prevented?**

You can’t prevent congenital ptosis since it’s present at birth (your baby is born with it). Most causes of acquired ptosis are difficult to prevent, as well.

## **Outlook / Prognosis**

The outlook (prognosis) for ptosis depends on the type and severity of the condition. If you need surgery, the outlook is generally good. Most surgeries to correct drooping eyelids are very successful.

## **Living With**

### **When should I see my healthcare provider?**

You should see your eye care specialist if ptosis is affecting your vision. You should also reach out to them if:

* The condition affects your appearance and it bothers you.
* One eyelid suddenly starts drooping or closing.
* You have any double vision or pain.

If your child has ptosis, they should see an eye care specialist right away to assess their condition. Sometimes, their provider may recommend treatment immediately to prevent issues with vision development. Regardless if they receive treatment right away or later on, they should see an ophthalmologist regularly to keep track of their vision through regular eye exams.

**Possible complications**

Complications can occur after ptosis surgery. Immediately following surgery, your eyelid height and shape may be asymmetrical. Your surgeon may have also under- or overcorrected your condition, which should resolve over time. Other complications of ptosis surgery include:

* Bleeding from the wound.
* Surgical site infection.
* Damage to your cornea.
* Incomplete or abnormal closure of your eyelids.
* Recurrent (returning) ptosis.

#### **Eye drops for ptosis**

A prescription eye drop medication for adults with some forms of acquired ptosis is available. The medication is called oxymetazoline, and it targets the levator muscle. After using the drops, some people have noticed their eyelid opens wider. To continue working, you must use the drops every day. The drops don’t work for all forms of ptosis, so talk to your ophthalmologist to see if they’ll work for you.

**Differential diagnosis**

Pseudoptosis is the false perception of the ptosis, which may result from the following:

* Contralateral retracted lid
* Downward deviated ipsilateral eyeball followed by the upper lid
* Overhanging skin over the upper lid
* Brow ptosis
* Upper lid swelling, i.e., preseptal cellulitis, orbital cellulitis, chalazion, etc.
* Volume deficit as seen in microphthalmos, phthisis bulbi, enophthalmos, etc

**Epidemiology data**

Among all cases of ptosis, congenital ptosis is the most common type which seems to be more prevalent in males. Simple congenital ptosis is the most prevalent form of congenital ptosis. Among acquired cases, aponeurotic ptosis is the most common type which usually presents in late adulthood. Enough data is not available yet, about the incidence of ptosis. However, the prevalence of ptosis does not seem to be affected by other epidemiological factors such as race, etc.

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**Refractive errors (myopia, hyperopia, astigmatism, presbyopia, hypermetropia)**

**Definition and description**

Refractive Errors: These encompasses a catalogue of diseases which affect the eye's ability to focus light correctly. These can be corrected with eyeglasses, contact lenses, or surgery. Some of the diseases that can be termed as “Refractive Errors” include:

A refractive error is something about the natural shape of your eyes that makes your vision blurry. Refractive errors are some of the most common vision problems people experience.

The parts of your eyes work together like a relay team to pass light that enters your eye along a pathway to your brain. Light passes through your cornea — the clear part at the front of your eye. Your cornea focuses that light through your lens into a signal that hits your retina — the layer at the very back of your eyeball. Your retina converts that light into electrical signals that your optic nerve sends to your brain. Your brain then uses those signals to create the images you see.

Depending on which type of refractive error you have, something about the shape of your eyeball, your cornea or your lens prevents your eyes from focusing correctly on objects you’re looking at. This distorts your vision.

There are a few types of refractive errors. Some make it hard to focus your eyes on objects close to you. Others make objects far away look blurry. No matter which type of refractive error you have, an eye care specialist can diagnose and treat it. They’ll check your eyes and vision and prescribe a treatment to help you see clearly again.

Most people with refractive errors develop them over time as they grow and develop. Many refractive errors first appear in kids. But you can develop a refractive error at any point in your life, even if you’ve had clear vision before.

Visit an eye care specialist as soon as you notice your vision changing or getting worse. Even if you already wear corrective lenses like eyeglasses or contact lenses, you need your eyes examined regularly.

#### **What are the types of refractive errors?**

The four most common types of refractive errors include:

**Myopia (nearsightedness)**

Definition and description

People who have nearsightedness (myopia) have difficulty seeing distant objects, but can see objects that are close to them clearly. For example, if you have nearsightedness, you might not be able to make out signs on the side of the road until they’re just a few feet away or right outside of your car’s window.

Usually, nearsightedness happens because your eye grows too long from front to back. Sometimes, nearsightedness happens because your cornea or lens is too curved.

Nearsightedness usually develops in kids when they’re around 10 years old.

Myopia is common. According to one estimate, more than 40% of people in the U.S. have nearsightedness. This number is rapidly rising, especially among school-aged children. Eye experts expect this trend to continue in the coming decades.

One in four parents has a child with some degree of nearsightedness. Some eye experts believe that if your child spends a great deal of time engaged in “near” activities, such as reading or using smartphones and computers, it may raise their risk of developing myopia.

#### **Types of myopia**

Eye specialists divide myopia broadly into simple myopia and pathologic myopia. Pathologic myopia is a newer name for degenerative myopia.

People with simple myopia have contact lenses or eyeglasses that help provide clear vision, while those with pathologic myopia may not be able to have clear vision even with corrective lenses.

## **Symptoms and Causes**

Focus that happens in front of the retina of your eye instead of at the retina results in myopia (nearsightedness).

### **What are the symptoms of myopia?**

If you have nearsightedness, you may notice:

* Faraway objects look blurred or fuzzy.
* Close items appear clear.
* Headaches.
* Eye strain.
* Squinting.
* Tiredness when driving, playing sports or looking more than a few feet away.

Some additional symptoms of myopia to watch for in your children include:

* Poor performance in school.
* Shortened attention span.
* Holding objects close to their face.

Most cases of myopia are mild and easily managed with eyeglasses, contact lenses or refractive surgery.

### **What causes myopia?**

If you have myopia, more than likely, at least one or both of your biological parents do, too. Eye experts are still unsure of the exact cause of myopia, but believe it to be a mix of hereditary and environmental factors.

It’s possible that you can inherit the ability to be myopic. If your lifestyle produces just the right conditions, you’ll develop it. For example, if you use your eyes for a lot of close-up work, like reading or working on a computer, you may develop myopia.

Myopia usually appears in childhood. Typically, the condition can worsen in early childhood but tends to level off by the end of teen years.

Because the light coming into your eyes doesn’t focus correctly, images are unclear. Think of it as being a little like a misdirected spotlight. If you shine a spotlight on the incorrect place in the distance, you won’t be able to see the correct object clearly.

#### **What are the risk factors for myopia?**

Risk factors for nearsightedness may include:

* A family history of myopia.
* Spending a lot of time doing “close-up” work, like reading or using screens like those on smartphones or computers.
* Not spending a lot of time outdoors. Certain studies indicate that this may be a factor in developing myopia.
* Ethnicity. Some groups of people have higher rates of myopia than others.

### **What are the complications of myopia?**

In most cases, providers can treat nearsightedness with glasses, contact lenses or corrective surgery, like LASIK. However, some cases of pathologic myopia can lead to more serious eye conditions, including:

* Cataracts.
* Glaucoma.
* Optic neuropathy.
* Neovascularization.
* Retinal detachment.

Pathologic myopia may make you more vulnerable to other more serious eye conditions. These include:

* Developing unwanted blood vessels in your eye (neovascularization).
* Glaucoma.
* Myopic optic neuropathy.
* Retinal detachment.
* Cataracts.

High myopia happens when your child’s eyeballs are too long, or their corneas are too steep.

## **Diagnosis and Tests**

An eye care provider can diagnose myopia using standard eye exams. Providers usually diagnose myopia in childhood, but it can also develop in adults because of visual stress or diabetes.

#### **Testing an adult for myopia**

Your provider will evaluate how your eyes focus light and measure the power of any corrective lenses you may need. First, they’ll test your visual acuity (sharpness) by asking you to read letters on an eye chart. Then, they’ll use a lighted retinoscope to measure how your retina reflects light.

Your provider may also use a phoropter. A phoropter is an instrument that measures the amount of your refractive error by placing a series of lenses in front of your eyes. This is how your provider measures the lens strength you need.

#### **Testing your child for myopia**

Your pediatrician will check your child’s eyes at each well-child visit. A first eye exam should be before age 1, if possible. If your child has no evident eye problems, then schedule a repeat eye exam before kindergarten.

As myopia runs in families, if your child has family members with vision issues, it’s even more important to test their eyes early. If you or your pediatrician notice any vision issues, your child may be referred to an optometrist or pediatric ophthalmologist.

During a children’s eye exam, your eye care provider will do a physical examination of your child’s eyes and check for a regular light reflex. For children between the ages of 3 and 5 years, your provider will also conduct vision screenings using eye chart tests, pictures, letters or the “tumbling E game,” also called the “Random E’s Visual Acuity Test.”

As your child’s vision continues to change as they grow, continue to make sure they get vision screenings by their pediatrician or eye care provider before first grade and every two years thereafter. While most schools conduct eye screenings, they’re usually not complete enough to diagnose myopia. Providers diagnose most children when they’re between the ages of 3 and 12.

Your provider may mention categories — mild, moderate or high myopia. These terms refer to the degree of nearsightedness as measured by refractive error. Refractive errors are issues with the natural shape of your eyes that make your vision blurry. It’s possible to have myopia and another refractive error, like astigmatism.

## **Management and Treatment**

Glasses or contact lenses can correct myopia in children and adults. For adults only (with rare exceptions for children), there are several types of refractive surgeries that can also correct myopia.

With myopia, your prescription for glasses or contact lenses is a negative number, such as -3.00. The higher the number, the stronger your lenses will be. The prescription helps your eye focus light on your retina, clearing up your distance vision.

* Eyeglasses: The most popular way for most people to correct myopia is with eyeglasses. Depending on the degree of vision correction needed, you’ll wear eyeglasses either daily or only when you need distance vision. You may only need glasses for driving. Some kids with myopia may only need glasses to play ball, watch a movie or view the chalkboard. Some people may need to wear glasses constantly to see clearly. A single-vision lens will make distance vision clearer. But people over 40 who have myopia may require a bifocal or progressive lens to see clearly both near and far.
* Contact lenses: Some people find that their distance vision is sharper and wider with contact lenses. A potential downside is they require more care to keep clean. Ask your provider which type might be right for your myopia level and other refractive errors.
* Ortho-k or CRT: Some people with mild myopia may be candidates for temporary corneal refractive contact lenses that you wear to bed to reshape your cornea temporarily, long enough to see for your daily activities.
* LASIK is a laser-assisted in situ keratomileus procedure, the most common surgery to correct nearsightedness. In a LASIK procedure, your ophthalmologist uses a laser to cut a flap through the top of your cornea, reshape the inner corneal tissue and then drop the flap back into place.
* LASEK is a laser-assisted subepithelial keratectomy procedure. In a LASEK procedure, your ophthalmologist uses a laser to cut a flap through only the top layer (epithelium) of your cornea, reshape the outer layers, and then close the flap.
* PRK is short for “photorefractive keratectomy,” which is a type of laser eye surgery used to correct mild or moderate nearsightedness. It may also correct farsightedness and/or astigmatism. In a PRK procedure, your ophthalmologist cuts off the front surface of your cornea and uses a laser to reshape the surface, which flattens it and allows light rays to focus on your retina. Unlike LASIK, the ophthalmologist doesn’t cut a flap, and your cornea will regrow its top layer in one to two weeks. PRK is better for people with corneas that are thinner or have a rough surface because it disrupts less corneal tissue than a comparable LASIK surgery.
* Phakic intraocular lenses: These are an option for people who have high myopia or whose corneas are too thin for PRK or LASIK. Your provider places phakic intraocular lenses inside of your eye just in front of your natural lens.
* Intraocular lens implant: This allows your ophthalmologist to surgically insert a new lens in your eye, replacing your natural one. This procedure happens before a cataract develops.
* Vision therapy: This is an option if spasms of your focusing muscles cause myopia. You can strengthen the muscles through eye exercises and improve your focus. This treatment isn’t appropriate for everyone with myopia. After an eye exam, your ophthalmologist will let you know if it’s an option for you.

## **Outlook / Prognosis**

Myopia is a condition that doesn’t go away. Treatments include using glasses or contact lenses. You may be able to get surgery to correct your vision.

### **What is the outlook for myopia?**

The outlook for being nearsighted may differ depending on the type of myopia.

Usually, providers can treat simple myopia easily. In rare cases of high myopia or pathologic myopia, your outlook may be different.

High myopia usually stops getting worse between the ages of 20 and 30. You’ll still be able to get glasses or contact lenses or you may be able to have surgery.

High myopia may lead to pathologic myopia and the possibility of more serious sight conditions later in life. These complications can lead to loss of sight.

Regular eye exams are important for everyone but are especially if you have high myopia or pathologic myopia. You should follow the schedule set out by your eye care provider.

## **Prevention**

You can’t prevent myopia as it’s a condition that tends to run in families, but you may be able to lower your risk of nearsightedness in some ways.

#### **How can I lower my risk of developing myopia?**

Some eye experts believe that you may be able to decrease your or your child’s risk of developing myopia by getting enough time outside and limiting the amount of time spent in front of screens. You may also want to be mindful of the amount of time doing close work like reading or sewing.

## **Living With**

### **How can you prevent myopia from getting worse?**

Though there’s no cure for myopia, there are everyday steps you can take that can support your overall eye health. These days, it’s especially important to set limits for your children (and yourself) on activities that lead to eye strain.

Try these sight-saving tips:

* Limit time on digital devices.
* Take screen breaks to stretch your eye muscles.
* Don’t read or work in dim light.
* Go outdoors and wear sunglasses when you’re out.
* Wear protective eye gear for sports/hobbies.
* Stop smoking.
* Schedule regular eye exams.
* Ask your provider about atropine eye drops to slow progression.
* Ask your provider about dual-focus contact lenses to slow progression in kids.

### **Which foods should I eat to keep my eyes as healthy as possible?**

Everyone’s eyes rely on nutrients from the foods we eat to maintain vital eye tissues and functions. Nutrition is especially essential to your child’s vision as their eyes grow and develop. In addition to limiting caffeinated colas and other soft drinks, keep hydrated by drinking enough water.

Also, try to eat foods that are rich in:

* Vitamin A. You need enough of the antioxidant vitamin A in your diet to maintain the surface of your eyes and healthy vision. There are vitamin A-rich sources for every diet preference. Plant-based choices include vegetables like sweet potatoes, leafy green vegetables and carrots. Or choose animal-based foods, such as cheese, oily fish or liver.
* Vitamin C. The best foods for getting a daily dose of vitamin C are fruits and vegetables, including oranges, grapefruit, strawberries and broccoli.
* Lutein. Eat leafy green vegetables to make sure to get enough lutein, which helps your eyes filter harmful blue light that can damage retinas.

You can supplement your or your child’s diet with a multivitamin if you think you or they aren’t getting enough vitamins and minerals. Remember, though, that your body doesn’t absorb vitamins in pills as well as vitamins that occur naturally in foods. And it’s important to check with your healthcare provider before starting any supplements.

Taking safe care of your and your family’s vision means regular eye exams, a good eye care routine and a healthy diet. Keeping those healthy habits will help you all to see a future filled with all the things you love.

### **When should I see a doctor about myopia?**

Regular eye exams are important for everyone. It’s especially important to contact an eye care provider if you have any type of change in your vision.

If you have kids and you notice that they squint a lot or pull things close to their faces to see them, make an appointment.

In any case of extreme changes in vision — like a sudden loss of vision or noticing a significant increase in the number of floaters or flashes of light you see — get immediate medical help. Some conditions, like retinal detachment, are medical emergencies.

**Differential diagnosis**

## 1. True Myopia

* Characterized by excessive axial length of the eye or increased refractive power causing light to focus in front of the retina.
* Can be simple (physiological) or pathologic (degenerative myopia) with associated structural changes like posterior staphyloma and myopic maculopathy.

## 2. Pseudomyopia (Spasm of Accommodation)

* Transient myopic shift caused by ciliary muscle spasm.
* Often related to prolonged near work, eye strain, or certain medications.
* Refractive error fluctuates and improves with cycloplegic refraction.

## 3. Astigmatism

* Irregular corneal or lenticular curvature causing blurred vision.
* Can coexist with myopia or mimic it if the axis and magnitude produce similar symptoms.
* Regular astigmatism affects vertical/horizontal meridians; oblique astigmatism affects oblique meridians.

## 4. Keratoconus and Other Corneal Ectasias

* Progressive corneal thinning and protrusion causing irregular astigmatism and myopic shift.
* Early keratoconus may mimic or be confused with high myopia.
* Corneal topography and tomography help differentiate keratoconus from simple myopia.

## 5. Retinopathy of Prematurity (ROP) and Other Posterior Segment Abnormalities

* Conditions like ROP can cause increased axial length and myopic refractive error.
* Posterior staphyloma or scleral buckle surgery can induce axial elongation mimicking myopia.

## 6. Lens-Induced Myopia

* Changes in lens shape or position (e.g., cataract, diabetes-induced lens swelling) can cause a myopic shift.
* Usually occurs in adults and can be transient.

## Diagnostic Approach

* Refraction Testing: Objective and subjective refraction to quantify myopia.
* Cycloplegic Refraction: To differentiate true myopia from pseudomyopia.
* Corneal Topography/Tomography: To detect keratoconus or other corneal irregularities.
* Axial Length Measurement: To confirm elongation in pathologic myopia.
* Fundus Examination: To assess for retinal or posterior segment pathology.

**Epidemiology data**

The prevalence of myopia in children aged 5–17 years varies globally and is the highest in Asians (18.5%), followed by Hispanics (13.2%), African Americans (6.6%), and Caucasians (4.4%). A study reported 20 to 30% prevalence in children aged 6 to 7 years in Taiwan and Singapore and up to 84% in high school students in Taiwan

Investigators in China found an increasing prevalence, ranging from 5.7% in children aged 5 years to 78.1% in those aged 15 years. Furthermore, the prevalence is 49.7% and 37.2% in children aged 10 to 15 years in Sweden and Greece, respectively.The prevalence in adults aged >44 years in the United Kingdom is 49%. The high prevalence in adults has been attributed to the presence of the lenticular component

[Myopia (Nearsightedness): Causes, Symptoms & Treatment](https://my.clevelandclinic.org/health/diseases/8579-myopia-nearsightedness#overview)

[Myopia - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK580529/#article-142919.s4)

**Hyperopia/hypermetropia (farsightedness)**

**Definition and description**

Farsightedness (hyperopia) makes it hard to see objects close to you clearly. It’s the opposite of myopia. If you have farsightedness, you might be able to read words on a screen on the other side of the room clearly, but will struggle to read notes you’re typing on your laptop that’s right in front of you.

Your eye growing too short from front to back causes farsightedness. Another cause is your cornea or lens being too flat (not being curved enough).

People with farsightedness are usually born with it.

**Causes and risk factors**

* **The cornea** is the clear, dome-shaped front surface of your eye.
* **The lens** is a clear structure about the size and shape of an M&M's candy.

In a normally shaped eye, each of these focusing elements has a perfectly smooth curvature, like the surface of a marble. A cornea and lens with such curvature bend (refract) all incoming light to make a sharply focused image directly on the retina, at the back of your eye.

### **A refractive error**

If your cornea or lens isn't evenly and smoothly curved, light rays aren't refracted properly, and you have a refractive error.

Farsightedness occurs when your eyeball is shorter than normal or your cornea is curved too little. The effect is the opposite of nearsightedness.

### **Other refractive errors**

In addition to farsightedness, other refractive errors include:

* **Nearsightedness (myopia).** Nearsightedness usually occurs when your eyeball is longer than normal or your cornea is curved too steeply. Instead of being focused precisely on your retina, light is focused in front of your retina, resulting in a blurry appearance for distant objects.
* **Astigmatism.** This occurs when your cornea or lens is curved more steeply in one direction than it is in another. Uncorrected astigmatism blurs your vision.

**Signs and symptoms**

Farsightedness may mean:

* Nearby objects may appear blurry
* You need to squint to see clearly
* You have eyestrain, including burning eyes, and aching in or around the eyes
* You have general eye discomfort or a headache after doing close tasks, such as reading, writing, computer work or drawing, for a time

**Diagnosis methods**

Farsightedness is diagnosed by a basic eye exam, which includes a refraction assessment and an eye health exam.

A refraction assessment determines if you have vision problems such as nearsightedness or farsightedness, astigmatism, or presbyopia. Your doctor may use various instruments and ask you to look through several lenses to test your distance and close-up vision.

Your eye doctor likely will put drops in your eyes to dilate your pupils for the eye health exam. This may make your eyes more light sensitive for a few hours after the exam. Dilation enables your doctor to see wider views inside of your eyes.

**Treatment options**

The goal of treating farsightedness is to help focus light on the retina through the use of corrective lenses or refractive surgery.

### **Prescription lenses**

In young people, treatment isn't always necessary because the crystalline lenses inside the eyes are flexible enough to compensate for the condition. Depending on the degree of farsightedness, you may need prescription lenses to improve your near vision. This is especially likely as you age and the lenses inside your eyes become less flexible.

Wearing prescription lenses treats farsightedness by counteracting the decreased curvature of your cornea or the smaller size (length) of your eye. Types of prescription lenses include:

* **Eyeglasses.** This is a simple, safe way to sharpen vision caused by farsightedness. The variety of eyeglass lenses is wide and includes single vision, bifocals, trifocals and progressive multifocals.
* **Contact lenses.** These lenses are worn right on your eyes. They are available in a variety of materials and designs, including soft and rigid, gas permeable in combination with spherical, toric, multifocal and monovision designs. Ask your eye doctor about the pros and cons of contact lenses and what might be best for you.

### **Refractive surgery**

Although most refractive surgical procedures are used to treat nearsightedness, they can also be used for mild to moderate farsightedness. These surgical treatments correct farsightedness by reshaping the curvature of your cornea. Refractive surgery methods include:

* **Laser-assisted in situ keratomileusis (LASIK).** With this procedure, your eye surgeon makes a thin, hinged flap into your cornea. He or she then uses a laser to adjust the curves of the cornea that corrects the farsightedness. Recovery from LASIK surgery is usually more rapid and causes less discomfort than other corneal surgeries.
* **Laser-assisted subepithelial keratectomy (LASEK).** The surgeon creates an ultra-thin flap only in the cornea's outer protective cover (epithelium). He or she then uses a laser to reshape the cornea's outer layers, changing its curve, and then replaces the epithelium.
* **Photorefractive keratectomy (PRK).** This procedure is similar to LASEK, except the surgeon completely removes the epithelium, then uses the laser to reshape the cornea. The epithelium is not replaced, but will grow back naturally, conforming to your cornea's new shape.

## **Lifestyle and home remedies**

You can't prevent farsightedness, but you can help protect your eyes and your vision by following these tips:

* **Have your eyes checked.** Do this regularly even if you see well.
* **Control chronic health conditions.** Certain conditions, such as diabetes and high blood pressure, can affect your vision if not treated.
* **Protect your eyes from the sun.** Wear sunglasses that block ultraviolet (UV) radiation.
* **Prevent eye injuries.** Wear protective eyewear when doing certain things, such as playing sports, mowing the lawn, painting or using other products with toxic fumes.
* **Eat healthy foods.** Try to eat plenty of leafy greens, other vegetables and fruits. And studies show that your eyes benefit if you also include in your diet fish high in omega-3 fatty acids, such as tuna and salmon.
* **Don't smoke.** Just as smoking isn't good for the rest of your body, smoking can adversely affect your eye health as well.
* **Use the right corrective lenses.** The right lenses optimize your vision. Having regular exams will ensure that your prescription is correct.
* **Use good lighting.** Turn up or add light to see better.
* **Reduce eyestrain.** Look away from your computer or near-task work, including reading, every 20 minutes — for 20 seconds — at something 20 feet away.

See your doctor immediately if you have any of these symptoms: Sudden loss of vision in one eye with or without pain; sudden hazy or blurred vision; double vision; or visual flashes of light, black spots or halos around lights. This may represent a serious medical or eye condition.

**Prevention tips**

There’s no proven way to prevent hyperopia.

However, some lifestyle habits can help keep your eyes healthy. Tips include:

* Eat a nutritious diet. Nutrients like vitamin A, vitamin C, vitamin E and lutein help protect your vision. To get these benefits, add lots of fruit (like grapefruit and strawberries) and veggies (like leafy greens) to your plate.
* Get regular eye exams. Your provider can check for eye problems before you have symptoms.
* Wear sunglasses, even on cloudy days. Choose sunglasses that block 99% or more of the sun’s ultraviolet (UV) radiation.
* Rest your eyes regularly. Looking at a screen for hours can tire your eyes and lead to computer vision syndrome. Making some small changes to your routine can help prevent or ease discomfort.

**Prognosis**

Hyperopia (farsightedness) doesn’t go away unless you have surgery. But even after surgery, your vision can change over time. This is a natural part of aging.

Glasses or contacts can correct your vision and help your eyes focus. But when you’re not wearing them, you may have symptoms of hyperopia. Plus, your vision can still change and get blurrier over time. You may notice that your glasses don’t help as much as they used to.

It’s important to wear your glasses or contacts as often as your provider recommends. You should also have regular eye exams in case you need to change the strength of your lenses.

**Possible complications**

Farsightedness can be associated with several problems, such as:

* **Crossed eyes.** Some children with farsightedness may develop crossed eyes. Specially designed eyeglasses that correct for part or all of the farsightedness may treat this problem.
* **Reduced quality of life.** With uncorrected farsightedness, you might not be able to perform a task as well as you wish. And your limited vision may detract from your enjoyment of day-to-day activities.
* **Eyestrain.** Uncorrected farsightedness may cause you to squint or strain your eyes to maintain focus. This can lead to eyestrain and headaches.
* **Impaired safety.** Your own safety and that of others may be jeopardized if you have an uncorrected vision problem. This could be especially serious if you are driving a car or operating heavy equipment.
* **Financial burden.** The cost of corrective lenses, eye exams and medical treatments can add up, especially with a chronic condition such as farsightedness.

**When to see a doctor / red flag**

If your degree of farsightedness is pronounced enough that you can't perform a task as well as you wish, or if your quality of vision detracts from your enjoyment of activities, see an eye doctor. He or she can determine the degree of your farsightedness and advise you of options to correct your vision.

Since it may not always be readily apparent that you're having trouble with your vision, the American Academy of Ophthalmology recommends the following intervals for regular eye exams:

#### **Adults**

If you're at high risk of certain eye diseases, such as glaucoma, get a dilated eye exam every one to two years, starting at age 40.

If you don't wear glasses or contacts, have no symptoms of eye trouble, and are at a low risk of developing eye diseases, such as glaucoma, get an eye exam at the following intervals:

* An initial exam at 40
* Every two to four years between ages 40 and 54
* Every one to three years between ages 55 and 64
* Every one to two years beginning at age 65

If you wear glasses or contacts or you have a health condition that affects the eyes, such as diabetes, you'll likely need to have your eyes checked regularly. Ask your eye doctor how frequently you need to schedule your appointments. But, if you notice problems with your vision, schedule an appointment with your eye doctor as soon as possible, even if you've recently had an eye exam. Blurred vision, for example, may suggest you need a prescription change, or it could be a sign of another problem.

#### **Children and adolescents**

Children need to be screened for eye disease and have their vision tested by a pediatrician, an ophthalmologist, an optometrist or another trained screener at the following ages and intervals.

* Age 6 months
* Age 3 years
* Before first grade and every two years during school years, at well-child visits, or through school or public screenings

**Differential diagnosis**

## Presbyopia

* Age-related loss of accommodation causing difficulty focusing on near objects.
* Unlike hyperopia, presbyopia typically affects adults over 40 and results from decreased lens elasticity rather than axial length or corneal curvature abnormalities.
* Both cause near vision blur but differ in pathophysiology and age of onset.

## 2. Orbital Tumors

* Mass lesions in the orbit can cause proptosis or displacement of the globe, altering axial length and refractive status, sometimes mimicking hyperopia.
* May be associated with other signs like pain, diplopia, or vision loss.

## 3. Serous Retinal Elevation

* Conditions causing serous retinal detachment or elevation (e.g., central serous chorioretinopathy) can induce a hyperopic shift by altering the retinal contour.
* Usually accompanied by visual distortion or scotoma.

## 4. Posterior Scleritis

* Inflammation of the sclera can cause thickening and choroidal effusion, leading to transient hyperopic shifts.
* Often painful with redness and other signs of ocular inflammation.

## 5. Cataracts

* Early nuclear sclerosis can induce a myopic shift, but cortical or posterior subcapsular cataracts may cause variable refractive changes including hyperopia.
* Lens changes can alter refractive power and accommodation.

## 6. Post-Refractive Surgery Changes

* Surgeries like LASIK or PRK aimed at correcting myopia can sometimes cause overcorrection or hyperopic shifts.
* History of surgery is key to diagnosis.

## 7. Functional Hyperopia

* Due to paralysis or spasm of accommodation, often seen in children or caused by drugs (e.g., cycloplegics).
* May be transient and identifiable by cycloplegic refraction.

**Epidemiology data** .

## Global Prevalence

* Approximately 30.6% of adults worldwide have hyperopia, with notable regional differences: Africa shows the highest prevalence at about 38.6%, the Americas about 37.2%, and Europe the lowest at around 23.1%.
* Total estimated cases globally range between 85 to 95 million adults, with Asia having the largest absolute number of cases (700–750 million), though with a lower prevalence percentage (12–18%) due to high myopia rates in that region[1](https://www.contactlenses.co.uk/education/hyperopia-stats).

## Age Distribution

* In children, hyperopia prevalence decreases with age: about 8.4% at age six, dropping to 2–3% between ages 9 to 14, and approximately 1% by age 15.
* In adults, prevalence increases with age. For example, in the United States, hyperopia affects about 9.9% of people aged 20–39, rising to 12.7% in those aged 40–59, and reaching 14.9% in those 60 and older[1](https://www.contactlenses.co.uk/education/hyperopia-stats).
* Adults over 40 years tend to have hyperopia prevalence ranging from 30% to 50%, with Africa and some parts of the Americas showing the highest rates, partly due to limited access to eye care.

## Regional Variations

* Africa has the highest prevalence and also the highest proportion of untreated hyperopia (around 60%) due to limited optometry services.
* Europe shows lower prevalence (around 23.1%), with northern countries like Finland, Sweden, and Norway having slightly higher rates (~35%) possibly due to genetic factors.
* Asia shows lower hyperopia prevalence (12–18%) compared to other regions, with countries like Japan and South Korea having higher rates (~20%) in older adults, while China and India have low hyperopia but very high myopia prevalence.
* Latin America and Oceania report prevalence rates between 22% and 30%, with rising cases linked to aging populations and lifestyle changes.

## Trends and Influencing Factors

* The prevalence of hyperopia has remained relatively stable over recent decades, unlike myopia which has shown a significant increase.
* Genetic, ethnic, and environmental factors (such as lifestyle and near work) influence hyperopia distribution.
* Uncorrected hyperopia in children can lead to amblyopia and affect learning and development, underscoring the importance of early detection and correction[4](https://www.scirp.org/journal/paperinformation?paperid=113193)

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**ASTIGMATISM**

*Alternative names: Astigmatism is also called astigma, stigmatism (an antonym for astigmatism); The term “astigmatism” itself is derived from the Greek words “a-” (without) and “stigma” (point or mark), referring to the inability of the eye to focus light to a single point.*

**DEFINITION / DESCRIPTION**

Astigmatism (uh- STIG-muh-tiz-um) is a common and generally treatable imperfection or an irregular curvature of the cornea or lenses of the eye that causes blurred distance and near vision.

Astigmatism occurs when either the front surface of the eye (cornea) or the lens inside the eye has mismatched curves. Instead of having one curve like a rounded ball, the surface is egg-shaped. This causes blurred vision at all distances.

Astigmatism is often present at birth and may occur in combination with nearsightedness or farsightedness. Often, it's not pronounced enough to require corrective action. When it is, treatment options are corrective lenses or surgery.

##### **Astigmatism**

Astigmatism can make objects at any distance look blurry. Usually, eyes are round. If you have astigmatism, your eye is shaped like a football or the back of a spoon. This makes light that enters your eyes bend and distort more than it should.

Astigmatism can develop at any point in your life. Some people are born with it. Others don’t experience it until they’re adults.

**CAUSES**

The eye has two structures with curved surfaces that bend (refract) light onto the retina, which makes the images:

* The cornea, the clear front surface of the eye along with the tear film
* The lens, a clear structure inside the eye that changes shape to help the eye focus on near objects

In a perfectly shaped eye, each of these elements has a round curvature, like the surface of a smooth ball. A cornea or lens with such curvature bends (refracts) all incoming light equally to make a sharply focused image directly on the retina at the back of the eye.

**A refractive error**

If either the cornea or the lens is egg-shaped with two mismatched curves, light rays aren't bent the same, which means that two different images form. These two images overlap or combine and result in blurred vision. Astigmatism is a type of refractive error.

Astigmatism occurs when the cornea or lens is curved more steeply in one direction than in another. You have corneal astigmatism if your cornea has mismatched curves. You have lenticular astigmatism if your lens has mismatched curves.

Either type of astigmatism can cause blurred vision. Blurred vision may occur more in one direction: horizontally, vertically or diagonally.

Astigmatism may be present from birth, or it may develop after an eye injury, disease or surgery. Astigmatism isn't caused or made worse by reading in poor light, sitting too close to the television or squinting.

**Other refractive errors**

Astigmatism may occur in combination with other refractive errors, which include:

* **Nearsightedness (myopia).** This occurs when the cornea is curved too much or the eye is longer than usual. Instead of being focused precisely on the retina, light is focused in front of the retina, making distant objects seem blurry.
* **Farsightedness (hyperopia).** This occurs when the cornea is curved too little or the eye is shorter than usual. The effect is the opposite of nearsightedness. When the eye is in a relaxed state, light never comes to a focus at the back of the eye, making nearby objects seem blurry

**RISK FACTORS**

Risk factors for astigmatism include:

* **Genetics.** Astigmatism can be hereditary, meaning it's passed down from parents.
* **Eye injury.**
* **Keratoconus.** Astigmatism risk is higher in people who have keratoconus, an eye condition in which the cornea thins and bulges outward.
* **Eye surgery.** Astigmatism can happen after eye surgery.

**SIGNS / SYMPTOMS**

Signs and symptoms of astigmatism may include:

* Blurred or distorted vision
* Eyestrain or discomfort
* Headaches
* Difficulty with night vision
* Squinting

**DIAGNOSIS METHOD**

Astigmatism is diagnosed with an eye exam. A complete eye exam involves both a series of tests to check eye health and a refraction, which determines how the eyes bend light. Your eye doctor may use various instruments, aim bright lights directly at your eyes and ask you to look through several lenses. Your doctor uses these tests to examine different aspects of your eyes and vision and to determine the prescription needed to provide clear vision with eyeglasses or contact lenses.

**TREATMENT OPTIONS**

The goal of treating astigmatism is to improve vision clarity and eye comfort. Treatments are corrective lenses or refractive surgery.

**Corrective lenses**

Wearing corrective lenses treats astigmatism by counteracting uneven curvatures of your cornea or lens.

Types of corrective lenses include:

* **Eyeglasses.** Eyeglasses are made with lenses that help compensate for the uneven shape of the eye. The lenses make the light bend into the eye properly. Eyeglasses can also correct for other refractive errors, such as nearsightedness or farsightedness.
* **Contact lenses.** Like eyeglasses, contact lenses can correct most astigmatism. They are available in a variety of types and styles.

Wearing contact lenses for extended periods of time increases the risk of infection in the eye.

Ask your eye doctor about the pros and cons and risks of contact lenses and what might be best for you.

**Refractive surgery**

Refractive surgery improves vision and reduces the need for eyeglasses or contact lenses. An eye surgeon uses a laser beam to reshape the curves of the cornea, which corrects the refractive error. Before surgery, doctors will evaluate you and determine if you're a candidate for refractive surgery.

Types of refractive surgery for astigmatism include:

* **Laser-assisted in-situ keratomileusis (LASIK).** With this procedure, an eye surgeon makes a thin, hinged flap in the cornea. He or she uses an excimer laser to sculpt the shape of the cornea and then repositions the flap.
* **Laser-assisted subepithelial keratectomy (LASEK).** Instead of creating a flap in the cornea, the surgeon loosens the cornea's thin protective cover (epithelium) with a special alcohol. He or she uses an excimer laser to change the curvature of the cornea and then repositions the loosened epithelium.
* **Photorefractive keratectomy (PRK).** This procedure is similar to LASEK, except the surgeon removes the epithelium. It will grow back naturally, conforming to the cornea's new shape. You may need to wear a bandage contact lens for a few days after surgery.
* **Epi-LASIK.** This is a variation of LASEK. The surgeon uses a special mechanized blunt blade — instead of the alcohol — to separate a very thin sheet of epithelium. He or she then uses an excimer laser to reshape the cornea and repositions the epithelium.
* **Small-incision lenticule extraction (SMILE).** This newer type of refractive surgery reshapes the cornea by using a laser to make a lens-shaped bit of tissue (lenticule) below the cornea's surface. The lenticule is then removed through a very small incision. For now, the SMILE procedure is only approved for treating mild nearsightedness.

Other types of refractive surgeries include clear lens extraction and implantable contact lenses. There is no one best method for refractive surgery, and you should make a decision only after a complete evaluation and thorough discussion with your surgeon.

**PROGNOSIS**

The prognosis of cases with astigmatism is usually good if treated on time, as there are multiple options available to correct astigmatism. Untreated patients, especially during childhood, may result in a permanent reduction in visual acuity and amblyopia.

Astigmatism is subject to change with time and will require new glasses and contact lenses. Refractive correction can often eliminate or reduce astigmatism in most cases. Patients with keratoconus can develop loss of visual acuity due to high astigmatism; hence timely and regular screening is mandated.

**POSSIBLE COMPLICATIONS**

Some of the possible complications that can occur after refractive surgery include:

* Undercorrection or overcorrection of your initial problem
* Visual side effects, such as a halo or starburst appearing around lights
* Dry eye
* Infection
* Corneal scarring
* Rarely, vision loss
* Defective vision
* Distorted vision
* Amblyopia
* Polyopia
* Strabismus
* Contact lens-induced infective keratitis

Discuss the potential risks and benefits of these procedures with your eye doctor.

**WHEN TO SEE A DOCTOR**

See an eye doctor if your eye symptoms detract from your enjoyment of activities or interfere with your ability to perform everyday tasks. An eye doctor can determine whether you have astigmatism and, if so, to what degree. He or she can then advise you of your options to correct your vision.

**Children and adolescents**

Children may not realize their vision is blurry, so they need to be screened for eye disease and have their vision tested by a pediatrician, an ophthalmologist, an optometrist or another trained screener at the following ages and intervals.

* During the newborn period
* At well-child visits until they reach school age
* During school years, every 1 to 2 years at well-child visits, at the eye doctor, or through school or public screenings

**Differential diagnosis**

* Myopia
* Hypermetropia
* Presbyopia

**EPIDEMIOLOGY**

Astigmatism typically changes with age. In early childhood, from 0 to 4 years of age, the cornea is steep, there is a high degree of corneal astigmatism, and the most common axis is against the rule of astigmatism. In the age group aged 4 to 18 years, the cornea flattens, astigmatism reduces, and small degrees of with the rule astigmatism is common. From 18 to 40 years, the cornea remains stable, and a small degree of astigmatism is common. From 40 years onwards, the cornea again steepens, and there is a shift in corneal astigmatism toward the rule.

Further, astigmatism varies amongst different ethnic groups. An increased prevalence of the rule astigmatism has been noted among Native Americans. Harvey, Dobson, and Miller reported astigmatism of 1.00D or more among 42% of school children. Poor nutrition has been postulated as a cause of reduced corneal rigidity.

As a result of this, the pressure from the upper eyelid steepens the vertical cornea and flattens the horizontal cornea. Increased rates of change in astigmatism have been reported among Asian subjects. The tightness of the Asian eyelids and narrow palpebral fissures have been suggested as causes of the greater rates of astigmatism change. Kleisnstein et al. reported the prevalence of one or more diopters among 33.6% of Asian and 36.9% of Hispanic children.

A study from Brazil reported the prevalence of myopia to be 2.7%, with a high prevalence of astigmatism of 16% (1 D astigmatism). They found a predominance of against the rule of astigmatism. In another study by Fuller et al., a high incidence of WTR astigmatism was seen in a small population subgroup of Bangladeshi children residing in East London.

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**Presbyopia**

**Definition and description**

##### **Age-related farsightedness (presbyopia)**

Age-related farsightedness (presbyopia) is a specific type of farsightedness that develops as you get older. Just like the farsightedness that can affect anyone, presbyopia makes it hard to see things up close. It’s usually the reason people need reading glasses as they age.

Presbyopia develops when the lens of your eye becomes less flexible and can’t focus on objects as well as it used to. It usually develops in people older than 40.

**Causes and risk factors**

To form an image, your eye relies on the cornea and the lens to focus the light reflected from objects. The closer the object, the more the lens flexes.

* **The cornea** is the clear, dome-shaped front surface of your eye.
* **The lens** is a clear structure about the size and shape of an M&M's candy.
* **Both of these structures** bend (refract) light entering your eye to focus the image on the retina, located on the inside back wall of your eye.

The lens, unlike the cornea, is somewhat flexible and can change shape with the help of a circular muscle that surrounds it. When you look at something at a distance, the circular muscle relaxes. When you look at something nearby, the muscle constricts, allowing the relatively elastic lens to curve and change its focusing power.

Presbyopia is caused by a hardening of the lens of your eye, which occurs with aging. As your lens becomes less flexible, it can no longer change shape to focus on close-up images. As a result, these images appear out of focus.

**Risk factors**

Certain factors can make you more likely to develop presbyopia, including:

* **Age.** Age is the greatest risk factor for presbyopia. Almost everyone experiences some degree of presbyopia after age 40.
* **Other medical conditions.** Being farsighted or having certain diseases — such as diabetes, multiple sclerosis or cardiovascular diseases — can increase your risk of premature presbyopia, which is presbyopia in people younger than 40.
* **Drugs.** Certain drugs are associated with premature presbyopic symptoms, including antidepressants, antihistamines and diuretics.

**Signs and symptoms**

Presbyopia develops gradually. You may first notice these signs and symptoms after age 40:

* A tendency to hold reading material farther away to make the letters clearer
* Blurred vision at normal reading distance
* Eyestrain or headaches after reading or doing close-up work

You may notice these symptoms are worse if you are tired or are in an area with dim lighting.

### **When to see a doctor**

See an eye doctor if blurry close-up vision is keeping you from reading, doing close-up work or enjoying other normal activities. He or she can determine whether you have presbyopia and advise you of your options.

Seek immediate medical care if you:

* Have a sudden loss of vision in one eye with or without eye pain
* Experience sudden hazy or blurred vision
* See flashes of light, black spots or halos around lights
* Have double vision

**Diagnosis methods**

Presbyopia is diagnosed by a basic eye exam, which includes a refraction assessment and an eye health exam.

A refraction assessment determines if you have nearsightedness or farsightedness, astigmatism, or presbyopia. Your doctor may use various instruments and ask you to look through several lenses to test your distance and close-up vision.

Your eye doctor likely will put drops in your eyes to dilate your pupils for the eye health exam. This may make your eyes more light sensitive for a few hours after the exam. Dilation enables your doctor to more easily view the inside of your eyes.

The American Academy of Ophthalmology recommends that adults have a complete eye exam every:

* Five to 10 years under age 40
* Two to four years between ages 40 and 54
* One to three years between ages 55 and 64
* One to two years beginning at age 65

You may need more-frequent exams if you have risk factors for eye disease or you need glasses or contact lenses.

**Treatment options**

The goal of treatment is to compensate for the inability of your eyes to focus on nearby objects. Treatment options include wearing corrective eyeglasses (spectacle lenses) or contact lenses, undergoing refractive surgery, or getting lens implants for presbyopia.

### **Eyeglasses**

Eyeglasses are a simple, safe way to correct vision problems caused by presbyopia. You may be able to use over-the-counter (nonprescription) reading glasses if you had good, uncorrected vision before developing presbyopia. Ask your eye doctor if non prescription glasses are OK for you.

Most non prescription reading glasses range in power from +1.00 diopter (D) to +3.00 D. When selecting reading glasses:

* Try different powers until you find the magnification that allows you to read comfortably, starting with the lower powers
* Test each pair on reading material held at a comfortable distance

You'll need prescription lenses for presbyopia if over-the-counter glasses are inadequate or if you already require prescription corrective lenses for nearsightedness, farsightedness or astigmatism. Your choices include:

* **Prescription reading glasses.** If you have no other vision problems, you can use glasses with prescription lenses for reading only. You will need to remove these when you're not reading.
* **Bifocals.** These lenses have a visible horizontal line that separates your distance prescription, above the line, and your reading prescription, below the line.
* **Trifocals.** These glasses have corrections for close-up work, middle distance vision — such as for computer screens — and distance vision. Trifocals come with two visible horizontal lines in the lenses.
* **Progressive multifocals.** This type of lens has no visible horizontal lines, but has multiple powers for distance, middle distance and close-up corrections. Different areas of the lens have different focusing strengths.
* **Office progressives.** These lenses have corrections for computer-distance and close work. You generally use these at a computer or for reading and remove them for driving or walking around.

### **Contact lenses**

People who don't want to wear eyeglasses often try contact lenses to improve their vision problems caused by presbyopia. This option may not work for you if you have certain conditions related to your eyelids, tear ducts or the surfaces of your eyes such as dry eye.

Several lens types are available:

* **Bifocal contact lenses.** Bifocal contact lenses provide distance and close-up correction on each contact. In one type of bifocal lens, the bottom, reading portion of the lens is weighted to keep the lens correctly positioned on your eye. Newer types of bifocal contact lenses offer one type of correction through the edges (periphery) of each lens and the other type of correction through the center of the lenses.
* **Monovision contact lenses.** With monovision contacts, you wear a contact lens for distance vision in one eye (usually your dominant eye) and a contact lens for close-up vision in the other eye.
* **Modified monovision.** With this option, you wear a bifocal or multifocal contact lens in one eye and a contact lens set for distance in the other (usually your dominant eye). You use both eyes for distance and one eye for reading.

### **Refractive surgery**

Refractive surgery changes the shape of your cornea. For presbyopia, this treatment can be used to improve close-up vision in your nondominant eye. It's like wearing monovision contact lenses. Even after surgery, you may need to use eyeglasses for close-up work.

Talk with your doctor about the possible side effects, as this procedure is not reversible. You might want to try monovision contact lenses for a while before you commit to surgery.

Refractive surgical procedures include:

* **Conductive keratoplasty.** This procedure uses radiofrequency energy to apply heat to tiny spots around the cornea. The heat causes the edge of the cornea to shrink slightly, increasing its curve (steepness) and focusing ability. The results of conductive keratoplasty are variable and may not be long lasting.
* **Laser-assisted in situ keratomileusis (LASIK).** With this procedure, your eye surgeon cuts a thin, hinged flap away from the front of your eye. He or she then uses a laser to remove inner layers of your cornea to steepen its domed shape.  
  Recovery from LASIK surgery is usually more rapid and less painful than other corneal surgeries.
* **Laser-assisted subepithelial keratectomy (LASEK).** The surgeon creates an ultra-thin flap only in the cornea's outer protective cover (epithelium). He or she then uses a laser to reshape the cornea's outer layers, steepening its curve, and then replaces the epithelium.
* **Photorefractive keratectomy (PRK).** This procedure is similar to LASEK, except the surgeon completely removes the epithelium, then uses the laser to reshape the cornea. The epithelium is not replaced, but will grow back naturally, conforming to your cornea's new shape.

### **Lens implants**

Some ophthalmologists use a procedure in which they remove the lens in each eye and replace it with a synthetic lens. This is called an intraocular lens.

Several types of lens implants are available for correcting presbyopia. Some allow your eye to see things both near and at a distance. Some change position or shape within the eye (accommodative lens). But lens implants can cause a decrease in the quality of your near vision, and you may still need reading glasses.

Possible side effects include glare and blurring. In addition, this surgery carries with it the same risks as those associated with cataract surgery, such as inflammation, infection, bleeding and glaucoma.

### **Corneal inlays**

Some people have had success with a presbyopia treatment that involves inserting a small plastic ring with a central opening, into the cornea of one eye. The opening acts like a pinhole camera and allows in focused light so that you can see close objects.

If you don't like the results of your corneal inlay procedure, your eye surgeon can remove the rings, leaving you free to consider other treatment options.

**Lifestyle and home remedies**

You can't prevent presbyopia. You can help protect your eyes and your vision by following these tips:

* **Have your eyes checked.** Do this regularly even if you see well.
* **Control chronic health conditions.** Certain conditions, such as diabetes and high blood pressure, can affect your vision if you don't receive proper treatment.
* **Protect your eyes from the sun.** Wear glasses or sunglasses that block ultraviolet (UV) radiation. This is especially important if you spend long hours in the sun or are taking a prescription medication that increases your sensitivity to UV radiation.
* **Prevent eye injuries.** Wear protective eyewear when doing certain things, such as playing sports, mowing the lawn, or painting or using other products with toxic fumes. Nonprescription reading glasses generally don't provide safety protection.
* **Eat healthy foods.** Try to eat plenty of fruits, leafy greens and other vegetables. These foods generally contain high levels of antioxidants as well as vitamin A and beta carotene. They're also vital to maintaining healthy vision.
* **Use the right glasses.** The right glasses optimize your vision. Having regular exams will ensure that your eyeglass prescription is correct.
* **Use good lighting.** Turn up or add light for better vision.
* **See your doctor immediately if you experience any of these symptoms** — sudden loss of vision in one eye with or without pain, sudden hazy or blurred vision, double vision, or see flashes of light, black spots or halos around lights. Any of these symptoms may signal a serious medical or eye condition.

**Prevention tips**

You can’t prevent presbyopia since it’s a part of the natural aging process. But you can take steps to protect your eye health. Tips include:

* Wear sunglasses to shield your eyes from harmful ultraviolet (UV) rays.
* Eat foods rich in nutrients that promote eye health, including vitamin A, vitamin C, vitamin E and lutein.
* Learn about computer vision syndrome and make adjustments to your routine to ease eye strain.

**Prognosis**

Talk to an eye care specialist about your symptoms and their impact on your daily life. They’ll help you find the most suitable corrective methods. You may need to try several different methods before finding one that works well for your needs and lifestyle.

**Differential diagnosis**

Presbyopia, a prevalent and anticipated age-related refractive disorder, must be carefully distinguished from other ocular illnesses and functional vision issues that may resemble its symptoms. The primary clinical characteristic of presbyopia is a gradual deterioration of near vision resulting from the diminished accommodating ability of the crystalline lens. However, many patients exhibit nonspecific symptoms, including eye strain, reading difficulties, blurred near vision, and headaches, all of which may potentially result from binocular vision problems, accommodative dysfunctions, early cataracts, or uncorrected refractive errors. Consequently, formulating a differential diagnosis is crucial for effective care and for preventing misdiagnosis, especially in younger or atypical patients.

Accommodative insufficiency, a condition not typically associated with age, commonly affects younger adults or adolescents and resembles presbyopia; however, it arises from a failure of the accommodative system rather than age-related changes in the lens. Individuals with accommodative insufficiency may encounter variable near vision, eye strain, and difficulty maintaining near tasks. In contrast to presbyopia, accommodative amplitude in these individuals can occasionally be enhanced with vision therapy, and the disease may be reversible. Binocular vision anomalies, including convergence insufficiency or excess, must be ruled out, particularly when patients experience diplopia or frontal headaches when reading. These diseases may simulate presbyopic symptoms and are generally diagnosed through cover testing, near point of convergence evaluation, and fusional reserve assessment.

Latent hyperopia is a significant factor in the differential diagnosis. In such cases, the patient may possess sufficient distant vision yet encounter strain during near tasks due to an undetected hyperopic refractive defect that necessitates excessive accommodative exertion. This condition can especially impact younger patients who retain accommodation reserves yet are starting to exhibit signs of visual fatigue. Cycloplegic refraction is crucial for identifying latent hyperopia, particularly in patients younger than 45 exhibiting early presbyopia symptoms. Inability to recognize this condition may lead to an incorrect diagnosis of presbyopia and inappropriate treatment.

Initial nuclear sclerotic cataracts may exhibit symptoms akin to presbyopia. Patients may experience a transition to myopia or, conversely, enhanced near vision, a condition referred to as second sight. In such instances, conventional refraction may erroneously indicate the resolution of presbyopia when the actual underlying condition is lenticular opacification. Slit-lamp biomicroscopy is crucial for identifying initial nuclear alterations that may not yet substantially affect visual acuity but signify the commencement of cataract development.

Dry eye syndrome and digital eye strain, both increasingly prevalent due to prolonged screen exposure, can cause near vision blurriness and discomfort that resemble presbyopia, particularly in individuals with significant visual requirements at intermediate and near distances.

Specific neurological disorders, such as progressive supranuclear palsy and cranial nerve palsies, can hinder accommodation or induce convergence impairments, leading to challenges with near vision. Although uncommon, these disorders should be considered when the clinical presentation deviates from age-related norms or when neurological symptoms are present. A comprehensive history and neurological evaluation are required in these instances..

Ocular conditions affecting near vision include:

* Macular and retinal diseases: In macular diseases, vision does not improve with refractive corrections, and optical coherence tomography of the retina may reveal the abnormalities.
* Diseases of the optic nerve
* Glaucoma
* Posterior subcapsular cataract: Unlike presbyopia, the near vision further deteriorates in bright light.
* Hypermetropia
* Astigmatism

**Epidemiology data** .

Presbyopia is a prevalent visual impairment affecting around 1.8 billion individuals worldwide. By 2030, this number is expected to increase to more than 2.1 billion.This condition is typical and physiological in individuals older than 50, although the precise age of start and severity differ among populations. Research indicates that presbyopia generally manifests symptomatically between the ages of 40 and 45, with nearly universal prevalence by age 60. Studies have shown that more than 80% of individuals aged 40 develop presbyopia. The worldwide prevalence of presbyopia is escalating as a result of an aging population and growing life expectancy. By 2050, more than 20% of the world's population is expected to be older than 60, which will have a significant impact on this condition in the near future. In numerous high-income nations, the percentage of adults older than 65 is increasing swiftly, resulting in a concomitant rise in the incidence of presbyopia. The ongoing increase in smartphone and digital screen usage highlights the need to manage presbyopia, given the heightened pressure on close vision tasks in both professional and recreational contexts.

The Global Burden of Disease Study indicates that uncorrected presbyopia leads to substantial productivity losses, estimated at over USD 25 billion worldwide, especially in low- and middle-income nations with restricted access to corrective glasses. Gender differences are prominent in specific populations. Systematic research showed that women experience presbyopia earlier than men and are less likely to utilize corrective services in low-income settings. This age-related alteration in accommodation affects both genders, but certain studies suggest an earlier onset in women, possibly due to hormonal and vocational disparities.The gender disparity may be ascribed to socioeconomic influences, cultural perspectives, and inequality in healthcare services.

Environmental factors such as temperature, humidity, and geographic latitude may significantly influence the onset and progression of presbyopia.Individuals residing in tropical climates often develop presbyopia at an early age, potentially due to increased exposure to UV radiation and various environmental stressors.Regional disparities in presbyopia correction are evident in the current literature. In wealthy nations such as the United States and certain regions of Europe, more than 60% of individuals with presbyopia use some correction for this condition. Conversely, in numerous low-resource areas, less than 20% of individuals with presbyopia have access to adequate visual aids. This gap highlights the necessity for public health initiatives focused on enhancing access to affordable eye care and presbyopic correction, particularly among aging populations in underdeveloped regions.

[Presbyopia - Diagnosis and treatment - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/presbyopia/diagnosis-treatment/drc-20363329)

[Presbyopia: Symptoms, Causes & Treatment](https://my.clevelandclinic.org/health/diseases/8577-presbyopia#outlook-prognosis)

## **Symptoms and Causes**

### **What are the symptoms of a refractive error?**

All refractive errors will make your vision worse. In addition to having trouble seeing clearly, your symptoms might include:

* Blurry vision either up close, at a distance or both.
* Double vision (diplopia).
* Headaches.
* Squinting.
* Eye strain.
* Eye pain.

Sometimes, kids might not know their vision is getting worse. If you notice your child has a hard time concentrating or if their grades in school suddenly get worse, they might have a refractive error. Visit your pediatrician or eye care specialist if you notice any changes in your child’s eyes, vision or behavior.

### **What causes refractive errors?**

Changes in the shape of your cornea, your lens or your whole eye can cause refractive errors. Which type of refractive error you have depends on how your eye is shaped.

They can also develop as you get older. Aging eyes can develop a refractive error you didn’t have when you were younger.

Some people develop a refractive error after surgery to remove cataracts from their eyes.

## **Diagnosis and Tests**

### **How are refractive errors diagnosed?**

An eye care specialist will diagnose refractive errors with an eye exam. They’ll look at your eyes (including inside them). They’ll also have you perform a visual acuity test. This will help determine which type of refractive error you have and how much it’s affecting your vision.

## **Management and Treatment**

### **How are refractive errors treated?**

Treatments for refractive errors include:

* Eyeglasses.
* Contact lenses.
* Vision correction surgery such as LASIK and photorefractive keratectomy (PRK).

Usually, your eye care specialist will prescribe you glasses or contacts before you have vision correction surgery. However, you might be a good candidate for vision correction surgery right away. Talk to your eye care specialist about which treatment will work the best for you.

## **Prevention**

### **How can I reduce my risk of developing a refractive error?**

There’s usually nothing you can do to prevent a refractive error from developing in your eyes. Because they’re caused by the shape of your eye, cornea or lens — or changes to these parts of your eyes — there’s no way to prevent refractive errors. There’s also nothing you can do to prevent your child from being born with a refractive error.

## **Outlook / Prognosis**

### **What can I expect if I have a refractive error?**

Having a refractive error might require you to wear glasses or contacts — or need vision correction surgery — but it shouldn’t impact your health.

The prescription (or strength) of your corrective lenses might change over time. Some people who receive vision correction surgery experience regression — the effects of the surgery disappearing over time.

If you have a refractive error, you have an increased risk of developing other conditions that affect your eyes, including:

* Glaucoma.
* Lazy eye (amblyopia).
* Crossed eyes (strabismus).
* Low vision.

## **Living With**

### **When should I have my eyes examined?**

Having your eyes and vision checked regularly can help your eye care specialist identify problems right away. How often you should get your eyes checked usually depends on your age:

* Kids: Your child’s pediatrician should check their eyes around the time they learn the alphabet, and then every one to two years.
* Adults younger than 40: Every five to 10 years.
* Adults between 40 and 54: Every two to four years.
* Adults older than 55: Every one to three years.

You might need your eyes checked more often than this if you wear glasses, contacts or need another type of visual aid. People with diabetes need their eyes checked more often than what’s listed here.

Ask your eye care specialist how often you need an eye exam.

### **When should I see my healthcare provider?**

See your healthcare provider or eye care specialist as soon as you notice any changes in your eyes or vision.

Go to the emergency room if you have any of the following symptoms:

* A sudden loss of vision.
* Severe eye pain.
* You see new flashes or floaters in your eyes.

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<https://my.clevelandclinic.org/health/diseases/24224-refractive-errors>

**Retinal detachment**

**Definition and description**

Retinal Detachment: Separation of the retina from its underlying tissue, a serious condition that can cause permanent vision loss if not treated promptly.

happens when the thin layer of tissue at the back of the eye pulls away from its regular position. This layer of tissue is called the retina. Retinal detachment is an emergency.

Retinal detachment separates the retinal cells from the layer of blood vessels that provides oxygen and nourishment to the eye. The longer retinal detachment goes without treatment, the greater the risk of permanent vision loss in the affected eye.

Symptoms of retinal detachment can include the following: reduced vision, the sudden appearance of dark floating shapes and flashes of light in your vision, and loss of side vision. Contacting an eye doctor, called an ophthalmologist, right away can help save your vision.

**Causes**

There are three main types of retinal detachment, and their causes vary:

* **Rhegmatogenous (reg-mu-TOJ-uh-nus).** This type of retinal detachment is the most common. A rhegmatogenous detachment is caused by a hole or tear in the retina that lets fluid pass through and collect underneath the retina. This fluid builds up and causes the retina to pull away from underlying tissues. The areas where the retina detaches lose their blood supply and stop working. This causes you to lose vision.

The most common cause of rhegmatogenous detachment is aging. As you age, gel-like material that fills the inside of your eye, called vitreous (VIT-ree-us), may change in texture and shrink or become more liquid. Usually, the vitreous separates from the surface of the retina without any complications. This is a common condition called a posterior vitreous detachment (PVD).

As the vitreous separates or peels off the retina, it may tug on the retina with enough force to create a tear. Most of the time it doesn't. But if a PVD causes a tear and the tear isn't treated, the liquid vitreous can pass through the tear into the space behind the retina. This causes the retina to detach.

* **Tractional.** This type of detachment can happen when scar tissue grows on the retina's surface. The scar tissue causes the retina to pull away from the back of the eye. Tractional detachment usually is seen in people who have poorly controlled diabetes.
* **Exudative.** In this type of detachment, fluid builds up beneath the retina, but there are no holes or tears in the retina. Exudative detachment can be caused by age-related macular degeneration, infection, tumors or inflammatory conditions.

**Risk factors**

The following factors raise your risk of retinal detachment:

* Aging — retinal detachment is more common in people ages 40 to 70.
* Past retinal detachment in one eye.
* Family history of retinal detachment.
* Extreme nearsightedness, also called myopia.
* Past eye surgery, such as cataract removal.
* Past severe eye injury.
* History of other eye diseases or conditions, including retinoschisis, uveitis or thinning of the peripheral retina called lattice degeneration.

**Symptoms**

Retinal detachment is painless. Often, symptoms are present before a retinal detachment happens or before it has gotten worse. You may notice symptoms such as:

* The sudden appearance of tiny specks or squiggly lines that seem to drift through your field of vision. These are called floaters.
* Flashes of light in one or both eyes. These are called photopsias.
* Blurred vision.
* Side vision, also called peripheral vision, becomes worse.
* A curtain like a shadow over your field of vision.

**Diagnosis**

Diagnosis involves the steps that your healthcare professional takes to find out if retinal detachment is the cause of your symptoms. Your healthcare team may use the following tests and instruments to diagnose retinal detachment:

* **Retinal exam.** Your healthcare professional may use an instrument with a bright light and special lenses to check the back of your eye, including the retina. This type of device provides a detailed view of your whole eye. It lets your healthcare professional see any retinal holes, tears or detachments.
* **Ultrasound imaging.** Your healthcare professional may use this test if bleeding has happened in your eye. Bleeding makes it hard to see the retina.

Your healthcare professional likely will check both eyes even if you have symptoms in just one. If a retinal tear is not found at this visit, your healthcare professional may ask you to return within a few weeks. The return visit is done to confirm that your eye has not developed a delayed retinal tear due to the same vitreous detachment. Also, if you have new symptoms, it's important to return to your healthcare professional right away.

**Treatment**

Surgery is almost always the type of treatment used to repair a retinal tear, hole or detachment. Various techniques are available. Ask your ophthalmologist about the risks and benefits of your treatment options. Together you can decide what treatment or combination of treatments is best for you.

**Retinal tears**

When the retina has a tear or hole but hasn't yet become detached, your eye surgeon may suggest one of the following treatments. These treatments can help prevent retinal detachment and preserve vision.

* **Laser surgery, also called laser photocoagulation or retinopexy.** The surgeon directs a laser beam into the eye through the pupil. The laser burns around the retinal tear to create scarring that usually "welds" the retina to underlying tissue.
* **Freezing, also called cryopexy.** Before treatment starts, you're given medicine to numb your eye. Then the surgeon applies a freezing probe to the outer surface of the eye directly over the tear. The freezing causes a scar that helps secure the retina to the eye wall.

Both treatments can be done in the eye doctor's office. Most often, you can go home afterward. You'll likely be told not to do activities that might jar the eyes — such as running — for a couple of weeks or so.

If your retina has detached, you'll need surgery to repair it. It's ideal to get surgery within days of finding out that your retina has detached. The type of surgery that your surgeon recommends depends on factors such as the location of the retinal detachment and how severe it is.

* **Injecting air or gas into the eye.** This surgery is called pneumatic retinopexy (RET-ih-no-pek-see). A surgeon injects a bubble of air or gas into the center part of the eye, also called the vitreous cavity. When positioned properly, the bubble pushes the area of the retina that contains the hole or holes against the wall of the eye. This stops the flow of fluid into the space behind the retina. The surgeon also uses cryopexy or laser photocoagulation during the treatment to create scarring around the retinal break.

Fluid that had collected underneath the retina is absorbed by itself, and the retina can then stick to the wall of the eye. You may need to hold your head in a certain position for up to a week to keep the bubble in the proper position. The bubble goes away on its own in time.

* **Indenting the surface of the eye.** This surgery is called scleral (SKLAIR-ul) buckling. It involves the surgeon sewing a piece of silicone to the white part of the eye, called the sclera, over the affected area. This surgery indents the wall of the eye and relieves some of the force caused by the vitreous tugging on the retina. The silicone is placed in a way that doesn't block your vision, and it usually remains in place for life. During surgery, cryoretinopexy or laser photocoagulation may be done to help seal tears in the retina. If fluid has collected below the retina, the surgeon may drain it.
* **Draining and replacing the fluid in the eye.** This surgery is known as vitrectomy (vih-TREK-tuh-me). The surgeon removes the vitreous along with any tissue that is tugging on the retina. Air, gas or silicone oil is then injected into the vitreous space to help flatten the retina. During surgery, tears in the retina can be sealed with cryoretinopexy or laser photocoagulation. There may be fluid below the retina that needs to be drained.

The air or gas that is injected into the vitreous space is absorbed in time. The vitreous space refills with fluid. If silicone oil was used, it may be removed with surgery months later.

Vitrectomy may be combined with scleral buckling.

After surgery, your vision may take months to get better. You may need a second surgery for successful treatment. Some people never get back all of their lost vision.

**Prevention**

You can’t prevent rhegmatogenous retinal detachment, but you can take steps to lower your risk:

* **Get regular eye care:** Eye exams protect your eye health. If you have nearsightedness, eye exams are especially important. Myopia makes you more prone to retinal detachment. Your eye care provider should include dilated exams to find small retinal tears.
* **Stay safe:** Use safety goggles or other protection for your eyes when playing sports or doing other risky activities.
* **Get prompt treatment:** If you notice detached retina symptoms, see your eye care provider right away or go to the emergency room.
* **Maintain your overall health:** Manage chronic conditions, eat balanced meals and get regular exercise.

You can help to prevent diabetes-related tractional retinal detachment by improving your blood glucose levels and blood pressure.

**How often should I get regular eye exams?**

People who have an average risk of eye disease should get eye exams once a year. If you’re at higher risk for eye disease, you may need checkups more frequently. Talk to your provider to figure out your best exam schedule.

**Outlook / Prognosis**

**What can I expect if I have a retinal detachment?**

Your outlook depends on factors like how clear your vision was before the retinal detachment, how extensive your detachment was and if there are any other complicating factors. Your provider will talk to you about what type of vision improvement you can expect.

In general, surgery for rhegmatogenous retinal detachment is highly successful — the repair works about nine out of 10 times. Sometimes, people need more than one procedure to return the retina to its place.

**Can I get a detached retina again?**

It’s possible to get a detached retina more than once. You may need a second surgery if this happens. Talk to your provider about preventive steps you can take to protect your vision.

**Living With**

It’s essential that you follow the instructions you get from your eye care provider about positioning and about your activities.

Ask your provider for suggestions on how to make things easier, like using a firm neck pillow to help keep your head in place. If you must lie face down or stay in that position for most of your time, your provider’s office can help you get face-down equipment for your home.

**When to see a doctor**

See a healthcare professional right away if you have any symptoms of retinal detachment. This condition is an emergency that can cause lasting vision loss.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnoses for suspected retinal detachment include the following:

* Retinoschisis
* Choroidal effusion or serous choroidal detachment
* Choroidal mass
* Suprachoroidal hemorrhage (hemorrhagic choroidal detachment)

The clinician can differentiate the above with a dilated fundoscopic exam and appropriate imaging techniques. If the patient has a quadrantanopia or hemianopia in both eyes, a cerebrovascular event should be in the differential. RRD should be on the differential for patients who have a TRD.Massive subretinal hemorrhage from various causes, including choroidal neovascular membrane due to age-related macular degeneration, polypoidal choroidal vasculopathy, peripheral exudative hemorrhagic chorioretinopathy (PEHCR), blood dyscrasias, anticoagulants, and trauma cause hemorrhagic RD.

**EPIDEMIOLOGY**

The incidence and risk of RRD vary between study results; one showed 1 in 10,000 individuals, and another showed the annual risk of RRD to be about 6.3 to 17.9 per 100,000 individuals. Males may be at a slightly higher risk than females of getting an RRD. There may be a higher risk of RRD in those of Southeastern Asian descent compared to European White race persons, confounded by the fact that Southeastern Asian populations tended to have a higher risk of myopia and a longer axial length. Results from another study did not find a significant difference in risk factors, postoperative outcomes, and clinical features in patients with retinal detachments between Indian, Malay, and Chinese populations in Singapore

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**RETINOPATHY OF PREMATURITY**

**Definition and description**

Retinopathy of prematurity (ROP) is an eye condition that affects some infants who are born early (premature birth), particularly before 31 weeks. With ROP, abnormal blood vessels form in a baby’s retina. The retina is the layer of tissue at the back of your eye that converts light to electrical signals, which travel to your brain. Your brain processes these signals and creates the images that make up your vision.

The abnormal blood vessels that form in ROP usually cause no harm and require no special treatment other than monitoring. Up to 90% of babies with ROP get better without treatment and have normal vision. However, the condition can sometimes get worse and threaten a baby’s vision. In these cases, timely treatment is necessary to prevent permanent retinal damage and vision loss. Without treatment, advanced ROP can lead to blindness.

That’s why healthcare providers recommend screenings for at-risk babies soon after birth. These screenings check for signs of ROP and identify when a baby needs treatment. Your baby’s healthcare provider will tell you if your baby is at risk for ROP. They’ll also tell you when your baby needs screenings. It’s essential to follow the screening schedule they give you to lower your baby’s risk of serious vision problems.

In the U.S., about 14,000 to 16,000 infants develop ROP each year. About 90% of these babies have a mild form of ROP that doesn’t need treatment. About 1,100 to 1,500 have a severe form that needs treatment. ROP causes legal blindness in 400 to 600 infants per year.

## **Symptoms and Causes**

### **What are the signs and symptoms of retinopathy of prematurity?**

There are usually no obvious signs or symptoms that you can notice in your baby. An ophthalmologist needs to closely examine your baby’s eyes (including blood vessel formation in their retinas) to see if they have ROP.

### **What causes retinopathy of prematurity?**

Disruption to the normal process of blood vessel formation in your baby’s retinas causes ROP.

Your baby needs healthy retinas with normal blood supply to see the world around them. Retinal blood vessels develop throughout pregnancy but aren’t completely formed until close to birth. As a result, babies born prematurely don’t have fully formed blood vessels in their retinas. Those vessels continue to form after birth, but they may develop abnormally.

It’s not always possible to tell which babies will have ROP, but researchers know some factors raise a baby’s risk.

#### **Risk factors for retinopathy of prematurity**

Risk factors for ROP include:

* Preterm delivery before 31 weeks. The earlier the delivery, the higher the risk of ROP.
* Birth weight of 1,500 grams (about 3.3 pounds) or less.
* Respiratory distress syndrome.
* Intracranial hemorrhage (bleeding in the brain).
* Infections or other medical problems.

If your baby has one or more risk factors, their healthcare provider will recommend a screening soon after birth to check your baby’s eyes for signs of ROP.

### **What are the complications of this condition?**

Untreated, severe cases of retinopathy of prematurity can lead to retinal detachment. This means your baby’s retina pulls away from the supportive tissues around it. Retinal detachment can cause severe vision loss or blindness.

## **Diagnosis and Tests**

### **How is retinopathy of prematurity diagnosed?**

Neonatologists typically identify babies who are at risk for ROP. They refer these babies to an ophthalmologist for further evaluation. During this exam (also called a “screening”), an ophthalmologist uses eye drops to dilate your baby’s eyes and look for signs of ROP. They may take digital pictures of your baby’s retinas. This initial screening usually takes place four to six weeks after birth.

Different countries have different guidelines for which babies should be screened for ROP. In the U.S., infants are typically screened if they:

* Have a gestational age of 30 weeks or less.
* Have a birth weight of 1,500 grams (3.3 pounds) or less.
* Have a higher gestational age or birth weight but have other risk factors for ROP.

Your baby may need additional screenings every one to three weeks, or according to the timeline their provider gives you. Your baby’s ophthalmologist will tell you when they no longer need these exams. This is usually when the blood vessels in your baby’s retinas are fully formed and there’s no risk of retinal detachment.

If the ophthalmologist diagnoses your baby with ROP, they’ll use a staging system to identify the condition’s severity.

#### **Retinopathy of prematurity stages**

Ophthalmologists assign a stage to each case of ROP to help describe its severity and the need for treatment. These stages range from 1 to 5, with 5 being the most severe:

* Stages 1 and 2: Mild to moderate ROP that usually goes away without treatment.
* Stage 3: ROP that may need treatment to prevent retinal damage or detachment.
* Stage 4: Severe ROP that leads to partial retinal detachment and requires urgent treatment.
* Stage 5: Severe ROP that leads to total retinal detachment and requires urgent treatment. Vision loss or blindness may still result despite treatment.

Other terms you may hear include:

* Aggressive retinopathy of prematurity: A severe case of ROP that quickly gets worse.
* Plus disease: Severe ROP that includes the presence of widened (dilated) and wavy (tortuous) blood vessels in the retina.

## **Management and Treatment**

Treatment options for retinopathy of prematurity include:

* Laser therapy. This treatment creates a pattern of small burns on the outer edges of your baby’s retina. These burns prevent abnormal blood vessels from forming. Laser therapy successfully treats ROP about 90% of the time.
* Anti-VEGF therapy. This treatment involves injections (shots) into your baby’s eye to deliver medication that stops abnormal blood vessel growth.

Your baby’s ophthalmologist will tell you the pros and cons of these treatments and explain the most suitable treatment plan for your baby.

If your baby has a retinal detachment (stage 4 or 5 ROP), they’ll need further treatment, typically with a retina specialist. For example, the retinal specialist may recommend a surgery called a vitrectomy.

#### **When do you treat retinopathy of prematurity?**

Your baby needs treatment if they’re at risk for retinal detachment, or if retinal detachment has already occurred. Your baby’s ophthalmologist will determine the best timing for treatment based on the ROP stage and findings from screenings.

## **Prevention**

ROP happens due to premature birth. Therefore, any steps you can take to lower your risk of early delivery can help lower your baby’s chances of developing ROP. It’s important to seek medical care during pregnancy and follow your provider’s guidance.

It’s also important to remember that sometimes, premature birth can’t be avoided, even if you follow all available advice. If this happens, don’t blame yourself. Advances in treatments and technology help babies born prematurely to be healthy and have a good prognosis.

## **Outlook / Prognosis**

ROP often goes away on its own without permanent damage to your baby’s retina or vision. However, severe cases of ROP need treatment to prevent complications like retinal detachment and vision loss.

Talk to your baby’s ophthalmologist to learn what treatment they may need and how ROP may affect their vision in the future.

## **Living With**

The most important thing you can do is take your baby to all of the screening appointments that their ophthalmologist recommends. These screenings are vital for diagnosing and treating ROP quickly enough to lower the risk for permanent vision loss.

Babies who receive treatment for ROP need lifelong follow-up visits. These are especially important during early childhood. Your baby’s ophthalmologist will look for signs of abnormal blood vessel formation. This can happen despite successful treatment years prior.

Babies born prematurely who don’t have ROP also need regular eye exams. That’s because they face an increased risk of certain eye problems, including:

* Amblyopia (lazy eye).
* Strabismus (crossed eyes).
* Glaucoma.

Ask your baby’s ophthalmologist how often they should have eye exams, and stick to this schedule.

**Differential diagnosis**

* Familial exudative vitreoretinopathy
* Persistent fetal vasculature
* Incontinentia pigmenti

**Epidemiology**

Many important epidemiological risk factors for ROP were established in the Early Treatment for Retinopathy of Prematurity study. This randomized, prospective multicenter trial compared the safety of earlier vs. conventionally timed ablation of peripheral retina. The incidence of any stage of ROP was 68% among infants weighing less than 1251g. In the year 2010, a global number of 184,700 infants and 14.9 million preterm infants developed any stage ROP. Of those afflicted, 20,000 became blind or severely visually impaired, and 12,300 developed mild to moderate visual impairment.

The two strongest known risk factors for ROP are gestational age (GA) and birth weight (BW). A multicenter study of over 4000 infants with birthweight ≤1251g found that for each 100g increase in BW, odds of developing threshold ROP decreased by 27%, and for each extra week in GA, the odds of developing threshold ROP decreased by 19%.

Another important risk factor is oxygen. As mentioned above, the use of supplemental oxygen in combination with atmospheric oxygen results in the reversal of physiologic hypoxia, which then contributes to retinal ischemia and subsequent overgrowth of retinal vessels in ROP. Additionally, the concentration of oxygen delivered is an independent risk factor for ROP, whereby increased concentrations of O2 increase the risk of ROP. For every 12 hour period with transcutaneous PO2≥80mmHg, the risk of ROP doubles.Duration of oxygen therapy is a significant risk factor for severe ROP. Possible risk factors include hypertensive disorders of pregnancy, maternal diabetes, medication use, age, smoking, assisted conception, birth outside of a study center hospital, and multiple gestations.

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**RETINAL VEIN OCCLUSION**

*ALTERNATIVE NAMES:* Retinal Vein Occlusion (RVO) is also known as an “eye stroke”. It is sometimes known as “Rious Retinal Vascular Occlusions”

**DEFINITION / DESCRIPTION**

Retinal vein occlusion (RVO) is a partial or total blockage in a vein that drains blood from your retina. Your retina is a layer of tissue at the back of your eye that helps translate light into images you can see. A blockage in a retinal vein prevents blood from leaving your retina. This can lead to complications, including raised pressure in your eye and swelling. These issues need prompt treatment to prevent or minimize vision loss.

There’s no current safe way to unblock the vein. However, treatment can manage complications and protect your vision.

Eye care specialists tailor treatment to your individual needs. You may need multiple treatments ranging from injections to surgery to manage your condition.

**Types of retinal vein occlusion**

There are two types of RVO:

* **Central retinal vein occlusion (CRVO)**, or blockage of the main retinal vein.
* **Branch retinal vein occlusion (BRVO)**, or blockage of one of the smaller branch veins. This type is more common.

Retinal vein occlusion is the second most common disorder affecting your retina (diabetes-related retinopathy is the most common).

Researchers estimate that globally:

* Retinal vein occlusion affects over 16 million people.
* Central retinal vein occlusion affects between 1 and 4 in 1,000 people.
* Branch retinal vein occlusion affects between 6 and 12 in 1,000 people.

**CAUSES**

**What causes retinal vein occlusion?**

A disruption to normal blood flow through your retinal vein causes this condition. The disruption may happen due to:

* A blood clot.
* A slowdown of blood flow.
* Compression of your retinal vein at the point where it crosses paths with your retinal artery. Your retinal artery supplies oxygen-rich blood to your retina. Your retinal artery may grow stiff from aging or plaque buildup, and it may press on your retinal vein. This can damage the inner lining of your retinal vein, creating conditions where a blood clot is more likely to form.

**What are the risk factors for retinal vein occlusion?**

Being over age 40 is a major risk factor. RVO usually affects people in their 50s or 60s. However, this condition can also affect people younger than age 40. Having certain medical conditions can also raise your risk. These include:

* Atherosclerosis.
* Diabetes.
* Glaucoma.
* High blood pressure.

Prior history of retinal vein occlusion in one eye raises your risk of developing the condition in your other eye.

**SIGNS / SYMPTOMS**

Symptoms of retinal vein occlusion typically affect one eye and include:

* **Blurry vision or vision loss**: This may start suddenly or develop gradually over a period of hours or days.
* **Floaters**: These are dark spots or lines in your field of vision.
* **Pain or pressure in your eye**: This is typically in more severe cases.

You may not have any symptoms until complications arise. Some people don’t realize there’s a problem until their provider finds the issue during a routine eye exam.

**Diagnosis methods**

Eye care specialists diagnose RVO through an eye exam and retinal imaging tests. They also coordinate care with your primary care physician (PCP) to discover the cause of blood flow problems.

**Eye exam**

Your eye care specialist will dilate your pupils so they can see into the back of each eye. They’ll use a microscope and a head-mounted ophthalmoscope to shine a light into your eye. They’ll closely examine the inside of your eye to look for complications and signs of vision loss.

This exam can help:

* Distinguish between central and branch RVO.
* Identify signs of macular edema and abnormal blood vessel formation.
* Estimate how much of your retina lacks blood flow.

You may need further testing to diagnose RVO and show the extent of complications.

**Tests to diagnose retinal vein occlusion**

Your eye care specialist may use one or more of the following tests to help diagnose and describe your condition:

* **Fundus photography**: This form of retinal imaging shows the presence of abnormal new blood vessels and the amount of bleeding inside your eye.
* **Optical coherence tomography (OCT)**: This form of high-resolution imaging shows the presence of macular edema. It measures the thickness of your retina and provides precise numbers that help guide the treatment of your condition over time.
* **Fluorescein angiography**: For this test, your provider injects dye into a vein in your arm. The dye travels to the blood vessels in your retina and makes them stand out in imaging. Your provider may use this form of imaging to show the extent of a blockage in your retinal vein. This test also shows how much of your retina isn’t receiving adequate blood flow.

**Coordinated care**

Your eye care specialist and primary care physician will work together to find the cause of RVO and lower your risk for future issues. You may need blood tests to check your cholesterol levels, blood sugar and other important numbers.

**Management and Treatment**

There’s currently no way to reverse or cure the blockage in your retinal vein. But eye care specialists can prevent or treat the complications of retinal vein occlusion with:

* Anti-VEGF injections.
* Steroid injections.
* Pan retinal photocoagulation (PRP).
* Vitrectomy surgery.
* Medications to manage risk factors.

The goals of treatment are to:

* Improve your vision or prevent it from getting worse.
* Identify and treat complications that can harm your vision and eye health.
* Manage risk factors to prevent future problems.

Your provider will combine treatment options as necessary and explain the timing for each.

**Anti-VEGF injections**

This is a first line (first choice) treatment for people with macular edema. VEGF stands for vascular endothelial growth factor. This is a protein that spurs new blood vessel growth (angiogenesis). Too much VEGF can lead to the formation of abnormal blood vessels that can leak and cause swelling.

Anti-VEGF injections interrupt the production of VEGF in your eye to reduce swelling. Your provider gives you eye drops to numb your eye and reduce pain before injecting the medication into the gel-like substance (vitreous humor) that fills your eyeball. You may need injections at regular intervals for one to two years depending on your condition.

Specific medications you may receive in these injections include:

* Aflibercept.
* Bevacizumab.
* Ranibizumab.

**Steroid injections**

Injections of steroid medication into your eye can also help reduce swelling. However, in some people, steroid injections cause elevated eye pressure and cataracts. So, they’re often a second-line treatment when anti-VEGF injections aren’t adequate.

**Pan retinal photocoagulation (PRP)**

This laser surgery creates small burns in areas of your retina that lack blood flow. Doing so decreases the number of proteins (VEGF) that promote the formation of abnormal blood vessels. Reducing VEGF helps prevent neovascularization and related bleeding in your eye. It also helps keep your intraocular pressure stable.

**Vitrectomy surgery**

Posterior pars plana vitrectomy (PPV) is a surgery that helps people with retinal vein occlusion who have:

* Severe bleeding in their eye (vitreous hemorrhage).
* Bleeding that lasts more than four weeks.
* Bleeding that keeps coming back.
* Retinal detachment.

Surgery removes vitreous humor from your eye and repairs damage to your retina.

**Medications to manage risk factors**

Many people with retinal vein occlusion have underlying conditions like high blood pressure, diabetes or high cholesterol. These conditions can raise your risk of blood vessel problems. Your eye care specialist will work together with your primary care physician (PCP) to tailor treatment to your needs. Your PCP may prescribe medications to:

* Lower your blood pressure.
* Manage your cholesterol levels.
* Address other issues.

**Prevention**

Learning you’re at risk for retinal vein occlusion is the first step toward preventing it. Talk to your ophthalmologist or optometrist about your level of risk and how to lower it.

It’s also important to talk to your primary care physician about underlying conditions that raise your risk for blood flow problems. They’ll recommend treatments as needed to manage those conditions and help keep your eyes — and whole body — healthy.

Specific things you can do to lower your risk include:

* Follow a diet that supports your heart and blood vessel health.
* Make exercise part of your daily routine.
* Keep a weight that’s healthy for you.
* Avoid smoking and all tobacco products.

**Outlook / Prognosis**

**What can I expect if I have this condition?**

Your prognosis depends on many factors, including the location of the blockage and complications that arise. Some people have permanent vision damage, while others have vision that gradually gets better over time. Your eye care specialist is the best person to tell you exactly what you can expect in your individual situation.

Your provider may refer you to vision rehabilitation. This is a form of rehab that teaches you techniques for living with reduced vision. These may include using devices like magnifying glasses or assistive-computer technology. Your provider may also refer you to a social worker who can help you cope with lifestyle changes.

**Living With**

**How do I take care of myself?**

Living with retinal vein occlusion (RVO) can be stressful because you may need:

* Multiple eye injections
* Multiple laser treatments.
* Many follow-up appointments.
* Help getting to and from your appointments (if your condition or treatment prevents you from driving safely).

All of this may take a toll and feel overwhelming to you. Remember that your healthcare team is there to help you.

Talk to your providers about how you’re feeling. They may suggest resources to help you learn more about your condition and why all of this effort is so important. They may also connect you with support groups or other community resources where you can talk to people who are in a similar situation. Learning from others’ experiences and sharing your own can help make everything feel more manageable.

**When should I seek medical care?**

Your eye care specialist will tell you how often you need appointments for monitoring or treatment. Call them if you experience new or changing symptoms or have questions about your treatment plan.

**What questions should I ask my provider?**

You may want to ask your eye care specialist:

* What caused the blockage in my retinal vein?
* What treatments are best for me?
* What are the benefits and risks of each treatment?
* What follow-ups will I need?
* How will this condition affect my vision?
* What is my outlook?

**POSSIBLE COMPLICATIONS**

Retinal vein occlusion can lead to complications such as:

* **Cystoid macular edema**: This is swelling in the center of your retina (macula). It can cause blurry vision or loss of vision.
* **Neovascularization of the eye**: Abnormal blood vessels can form in different parts of your eye, typically your iris (rubeosis iridis). This happens in about 1 in 4 people with RVO. Abnormal blood vessels less commonly form in your retina.
* **Bleeding in your eye (vitreous hemorrhage)**: This is when blood leaks into your vitreous humor, the gel-like substance that fills your eyeball. It results from the formation of abnormal blood vessels, which are prone to leaking.
* **Neovascular glaucoma**: Abnormal blood vessels in your eye can cause pain and a dangerous increase in pressure inside your eye.
* **Retinal detachment**: Abnormal blood vessels in your retina may cause your retina to pull away from the tissues that support it.

People with RVO have a higher risk of cardiovascular diseases, including stroke, compared to people without RVO. This may be due to shared underlying risk factors like high blood pressure and atherosclerosis.

**DIFFERENTIAL DIAGNOSIS**

Important differential diagnosis of BRVO includes diabetic retinopathy and hypertensive retinopathy. Diabetic retinopathy can be discerned from BRVO based on a few key features. Diabetic retinopathy is usually bilateral whereas BRVO is usually unilateral. Although both BRVO and diabetic retinopathy may present with dot blot hemorrhages and microaneurysms, only in diabetic retinopathy will extend across the horizontal raphe. Hypertensive retinopathy is the same; hemorrhages are not confined to a single sector, and the findings are usually bilateral.

**EPIDEMIOLOGY**

Retinal venous occlusion (RVO) is the second most common retinal vascular pathology after diabetic retinopathy. The exact incidence of BRVO is difficult to gauge, given the frequent asymptomatic nature of BRVOs. Nonetheless, the Blue Mountain Eye study found a 10-year cumulative risk of RVO to be 1.6% in the United States while finding no predilection for gender or race. The Beaver Dam Eye Study found a 15-year cumulative risk of BRVO to be 1.8%, three times more than CRVO at 0.5%. Risk factors for developing BRVO include increasing age or age over 70 years old, history of systemic arterial hypertension, history of smoking, or a history of glaucoma. A pooled analysis of population-based studies from the United States found BRVO to have a higher prevalence in Asians and Hispanics compared to Caucasians, although this was not statistically significant. Developing BRVO in one eye increases the risk of BRVO in the fellow eye to 7% to 10%.

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**Retinitis pigmentosa**

**Definition and description**

Retinitis Pigmentosa is a genetic retinal degeneration causing night blindness and peripheral vision loss.

Retinitis pigmentosa (RP) is a group of rare eye diseases that affect the retina (the light-sensitive layer of tissue in the back of the eye). RP makes cells in the retina break down slowly over time, causing vision loss.

RP is a genetic disease that people are born with. Symptoms usually start in childhood, and most people eventually lose most of their sight.

There’s no cure for RP. But vision aids and rehabilitation (training) programs can help people with RP make the most of their vision.

The retina is a layer at the back of your eye that converts light into electrical signals, allowing your brain to see the world around you. Retinitis pigmentosa (RP) is the term for a group of inherited eye diseases (IRDs) that affect your retina. RP is the most common type of inherited eye disease. Examples of other inherited retinal diseases include:

* Cone-rod dystrophy.
* Congenital stationary night blindness.
* Leber congenital amaurosis.
* Usher syndrome.

Your eye is often compared to a non-digital camera. The front of your eye contains a lens (just like a lens of a camera). This lens focuses images on the inside of the back of your eye, the retina (just like film in a camera). If the camera film is damaged, it doesn’t matter how much you focus the camera, the picture is not going to be perfect.

Similarly, a diseased retina will affect your vision even if the front of your eye functions well. This includes people with corrective eyeglasses or contact lenses.

Your retina consists of special nerve cells that react to light. These cells include photoreceptor cells such as rods and cones and retinal pigment epithelium cells. For proper sight, it’s essential that these cells work together harmoniously. The genetic mutations that cause RP and other IRDs prevent these cells from functioning properly.

Since RP is a group of disorders, the visual changes vary among different people. Most people with RP have low vision, and some people go blind. The vision changes usually start in childhood. But, sometimes these changes occur so slowly that you don’t realize it’s happening. Some people have faster vision loss. In some types of RP, vision loss stops at a certain point.

Retinitis pigmentosa generally affects both eyes.

If you are diagnosed with RP or IRD, it’s important to obtain genetic testing that can sometimes determine the exact mutation causing the disease. Genetic testing is done with a specifically trained genetic counselor who can help with the ordering and interpretation of the test. Identifying the mutation is important because it can provide insight into how the disease may progress and how other family members may be affected. It also might qualify you to receive gene therapy or participate in a gene therapy clinical trial.

There are an estimated 1 in 3,500 to 1 in 4,000 people in Europe and the U.S. who have retinitis pigmentosa. Globally, RP affects about 1 in 3,000 to 1 in 4,000 people, or about two million people total. In the U.S., this total is estimated at about 100,000 people.

### **What causes retinitis pigmentosa?**

Retinitis pigmentosa, as other IRDs, is caused by changes in certain genes. These genes control the cells that make up your retina.

### **What are the signs and symptoms of retinitis pigmentosa?**

Early signs and symptoms of retinitis pigmentosa include:

* Problems with night vision.
* Problems seeing in dim light.
* Blind spots in peripheral (side) vision.

Later retinitis pigmentosa signs and symptoms may include:

* Having a sensation of twinkling or flashing light.
* Having tunnel vision (only central vision).
* Being sensitive to or uncomfortable in bright light (photophobia).
* Losing the ability to see color.
* Having very low vision.

**Diagnosis methods (tests, lab work, imaging, etc.)**

### **What tests will be done to diagnose retinitis pigmentosa?**

It’s important to schedule and keep regular eye examinations. If your healthcare provider thinks you may have retinitis pigmentosa, they will use some or all of the following tests to make their diagnosis. They may also suggest genetic testing and/or counseling.

#### **Dilated eye examination with visual field test**

You may get your eyes examined by an optometrist or an ophthalmologist. Your provider will:

* Ask you to read letters off a chart on the wall.
* Ask you to follow objects with your eyes.
* Test the pressure in your eyes.
* Check your side vision (peripheral vision) with a visual field test.
* Check how your pupils react to light.
* Dilate your pupils with eye drops so they can see inside your eye.
* Take images of the retina.

#### **Electroretinography (ERG) test**

Electroretinography is a test that measures your retina’s response to light. It checks the function of different retinal cells. Your healthcare provider will flash lights in front of your eyes to measure your retina’s activity. Electroretinography is a type of ophthalmic electrophysiology test. These types of tests can check how your eyes and brain process what you’re seeing by measuring the electrical activity in your retinas, optic nerves, and visual pathways in your brain.

#### **Optical coherence tomography scan**

Optical coherence tomography (OCT) is a noninvasive test that can measure the thickness of your retina and analyze the retinal integrity. You are asked to look at a target as a special camera takes an image of the back of your eye.

#### **Fundus autofluorescence test**

This type of imaging test is noninvasive and can reveal information about the health of your retinas. It’s used for diagnosis, treatment and monitoring.

## **Management and Treatment**

A lot of advances have been made in recent years in the field of RP and IRDs including the introduction of gene therapy.

The ways to manage RP include:

* Using low vision aids and assistive devices. There are a range of magnifiers and technology that can identify things or people that the wearer points to.
* Using sunglasses and other methods to avoid exposure to too much light. Light may make RP worse.
* Treating associated conditions, such as cystoid macular edema (CME), which may happen with RP. CME refers to collections of fluid in the middle of your retina.
* Treating cataracts, which happens when the lens of your eyes becomes cloudy, with surgery to remove them.

### **Are there other ways to treat retinitis pigmentosa?**

The FDA has approved voretigene neparvovec-ryzl (Luxturna®), a gene therapy product to treat a specific type of retinitis pigmentosa. People with mutations in both copies of the *RP65* gene may benefit from this type of therapy. This particular type of RP affects 1,000 to 2,000 people in the U.S.

There are ongoing clinical trials for gene therapy for the other types of RP and IRDs.

Some people with severe RP may be able to get an artificial retina, also called a retinal prosthesis.

## **Prevention**

### **How can I prevent retinitis pigmentosa?**

Because most forms of retinitis pigmentosa are inherited, you can’t prevent RP. You can, however, take steps to keep your eyes as healthy as possible by:

* Making and keeping regular appointments with your ophthalmologist.
* Wearing sunglasses and avoiding bright lights.
* Being as healthy as possible by eating right and exercising safely.

## **Prognosis**

## The prognosis for patients with retinitis pigmentosa depends on the age of onset and pattern of inheritance. Expect early-onset symptoms, severe vision loss, and night blindness with the autosomal recessive form of RP. The autosomal dominant expression is the least severe and is associated with the more gradual onset of symptoms later in adulthood. The most severe vision loss occurs with X-linked recessive RP. Tunnel vision is expected late in all forms of RP, and almost all patients with RP will be legally blind at some point in the progression of their disease. Fortunately, total vision loss is uncommon, as the macular function will generally allow light perception, even after acuity is lost. Many patients retain good central vision into their 40s or 50s. Genetic testing is now available to determine the specific gene mutation causing a patient's RP, which may help predict disease severity and progression.

## **Complications**

## Functional difficulties associated with RP include night blindness, progressive loss of vision, and a gradual reduction in peripheral and central vision. In advanced stages, patients with RP may become legally blind or have minimal vision. Severely impaired night vision may make navigating in low-light conditions challenging. RP typically leads to gradually losing peripheral vision (tunnel vision). This narrowing of the visual field can affect daily activities such as mobility and driving.

## In some cases, RP can progress to affect central vision, which is essential for tasks like reading and recognizing faces. Some individuals with RP may experience color vision abnormalities, including difficulty distinguishing between specific colors or a reduced ability to perceive colors accurately. Increased sensitivity to light (photophobia) can complicate RP. Bright lights can cause discomfort and glare, making it difficult to tolerate well-lit environments. RP can reduce contrast sensitivity, making distinguishing objects or text from their background difficult. Reduced peripheral vision and contrast sensitivity can lead to difficulties with depth perception, making it harder to judge distances accurately. Reduced vision, especially in low-light conditions, can increase the risk of accidents and injuries, both indoors and outdoors. Individuals with RP may have depression, anxiety, and reduced quality of life. As vision deteriorates, individuals with RP may become increasingly dependent on others for daily tasks and mobility, impacting their independence. Due to vision-related limitations, children and adults with RP may face difficulties in educational and occupational settings.

## Ocular complications associated with RP include cataracts, especially posterior subcapsular cataracts. The lens may have weak zonules resulting in anterior capsular phimosis, subluxation, or dislocation of capsular bag-intraocular lens complex after cataract surgery. Patients with RP may have retrolental or vitreous cells. Some cases are associated with intermediate uveitis and leaking cystoid macular edema on fluorescein angiography. Such patients may respond favorably with posterior subtenon triamcinolone acetonide, intravitreal, or systemic steroids.Macular complications associated with RP include foveoschisis (non-leaking on fluorescein angiogram), CME (petaloid leak on fluorescein angiogram), macular holes, epiretinal membrane, vitreomacular traction, and choroidal neovascular membrane. Myopia and astigmatism may be expected in patients with RP.Both open-angle and closed-angle glaucoma may be associated with RP. The prevalence of angle-closure glaucoma has been reported as 1% to 2.3% in populations of Canada and China, respectively. Around 2% to 12% of patients with RP have primary open-angle glaucoma. Around 68% of retinal detachments associated with RP are rhegmatogenous. There may be an absence of complete posterior vitreous detachment and a predominance of round holes in younger patients. Proliferative vitreoretinopathy is common, and titration of laser burns is necessary during surgery for rhegmatogenous retinal detachment. Coats disease-like exudative vitreoretinopathy may be noted in up to 5% of patients with RP. Such manifestation is usually bilateral and may cause exudative retinal detachment. Patients with RP may develop retinal vascular abnormalities like microaneurysms, telangiectasia, and neovascularization. These abnormalities can cause recurrent vitreous hemorrhage.

**Differential diagnosis**

The differential diagnosis for progressive vision loss is complex. RP is a clinical diagnosis confirmed by bilateral eye involvement with night vision disturbance and gradual loss of peripheral vision. Physical findings on fundoscopic examination reveal bone spicule pigmentation, with vascular narrowing and optic disc pallor. Note that there are variants of RP with significant differences in physical findings.

Retinitis punctata albescens (RPA, also called progressive RPA) is an autosomal recessive variant of RP that frequently presents as night blindness in childhood. Mutations in *RLBP1* usually cause this condition. On direct fundoscopic examination, findings include less prominent (or even absent) disc pallor, less pronounced vascular narrowing, and bony spicules are rare or absent altogether. Instead, small white spots cover the majority of the fundus in RPA. Awareness of such variants is necessary to avoid excluding RP and its subclasses from the differential. RPA is progressive with a progressive decline of visual fields and other visual parameters compared to fundus albipunctatus (congenital stationary night blindness), which is a stationary (non progressive) disease. However, a mutation in the *RLBP1* is known to be associated with Bothnia retinal dystrophy, fundus albipunctatus, and Newfoundland rod-cone dystrophy. Thus, there is a phenotypic variability.

Other diseases may mimic the early findings of this condition; careful evaluation is necessary to make the correct diagnosis. One possible mimic is cone-rod dystrophy. This progressive retinopathy can be differentiated from RP by the onset of dyschromatopsia before nyctalopia, as the cone photoreceptors are affected rather than the rods. Another consideration for nyctalopia in childhood is congenital stationary night blindness (CSNB). CSN presents with 2 types and nyctalopia in childhood. The first type is CSNB (both Riggs and Schubert-Bornstein type), which presents without abnormalities on the fundoscopic exam. The second type has visual fundoscopic abnormalities, including Oguchi disease and fundus albipunctatus. Oguchi disease is associated with the Mizuo-Nakamura phenomenon in which the fundus has a golden sheen on exposure to light, and the typical color of the fundus reappears after prolonged dark adaptation. Fundus albipunctata is characterized by small yellowish-white round dots sparing the fovea and extending to or beyond the mid-periphery. As previously mentioned, these conditions are nonprogressive.

Chorioretinal infections, such as syphilis, cytomegalovirus, or even Lyme disease, may produce symptoms suggestive of RP. Take a careful history to help identify these possibilities and perform laboratory testing for confirmation.

Vision loss can result from multiple other diseases, including sarcoidosis and systemic lupus erythematosus. Systemic involvement, in addition to vision loss, should prompt a thorough assessment of these diseases and other inflammatory processes. Also, consider trauma and Vitamin A deficiency, especially in children or adults with recent onset acquired nyctalopia. In older patients with recent onset of nyctalopia and retinal changes similar to RP, cancer-associated retinopathy and possible occult malignancy should be ruled out.

Unilateral retinitis pigmentosa (URP) is a rare retinitis pigmentosa that affects only one eye. According to results from multiple studies, URP diagnosis occurs only after ruling out all possible infective causes, and the affected eye shows all the clinical signs of retinitis pigmentosa. In contrast, the unaffected eye remains completely normal. However, the exact cause of most cases of this condition remains a mystery. Electroretinography and electrooculography help diagnose URP, as the affected eye will show greatly diminished rod function and non recordable or severely decreased electroretinogram responses, while the unaffected eye remains normal. Patients with URP often do not notice symptoms in the eye, though the condition can be present from birth. Some cases of URP appear to be genetic. Mukhopadhyay and colleagues described a patient with a germline mutation in the *RP1* gene who developed URP. However, the cause remains unclear of this patient's unilateral expression of the disease. Whether URP represents a distinct clinical entity or an abortive form of bilateral retinitis pigmentosa is still debated.

Diseases that can cause features similar to URP (called unilateral pseudo RP) or unilateral pigmentary retinopathy include

* **Inflammation:** diffuse unilateral subacute neuroretinitis (DUSN), acute zonal occult outer retinopathy (AZOOR), severe posterior uveitis, retinal vasculitis
* **Trauma:** blunt trauma (after resolution of commotion retinae or spontaneously settled retinal detachment), retained intraocular foreign body (iron foreign body-siderosis), forceps-associated birth trauma
* **Infection:** syphilis, cytomegalovirus, measles, rubella, toxoplasmosis, tuberculosis
* **Autoimmune disorders:** autoimmune retinopathy (AIR)
* **Malignancy:** cancer-associated retinopathy (CAR, paraneoplastic syndrome), choroidal melanoma
* **Drug toxicity:** chloroquine, thioridazine, phenothiazine, hydroxychloroquine, oral contraceptives, cephaloridine
* **Other retinal disorders:** self-settled retinal detachment
* **Vascular diseases:** ophthalmic artery occlusion

**Syndromic RP**

RP usually occurs as an isolated condition (nonsyndromic RP) but may be associated with several syndromes and genetic disorders. These syndromes often involve RP as one of their clinical features, along with other systemic manifestations. Important syndromes associated with RP include the following:

* **Usher syndrome:** Usher syndrome is the most common syndrome associated with RP. Around 18% of all patients with RP may have Usher syndrome. Sensorineural hearing loss and, in some cases, vestibular (balance) issues characterize this condition. There are 3 main types of Usher syndrome, each with different genetic causes and degrees of severity.
* **Bardet-Biedl syndrome:** Bardet-Biedl syndrome (BBS) is a rare genetic disorder characterized by RP, obesity, kidney dysfunction, extra fingers or toes (polydactyly), and other features such as intellectual disability and developmental delays. Multiple genes can be involved in BBS, leading to significant variability in clinical presentation.
* **Joubert Syndrome:** Joubert syndrome is a rare genetic disorder characterized by aplasia of the cerebellar vermis, causing the "molar tooth sign" on brain imaging and a range of neurological and systemic features. Some individuals with Joubert syndrome may also develop RP.
* **Senior-Løken Syndrome:** Senior-Løken syndrome combines the features of nephronophthisis and RP. This condition is a rare autosomal recessive disorder characterized by early-onset kidney disease and progressive vision loss.
* **Refsum Disease:** Refsum disease is a metabolic disorder characterized by the accumulation of phytanic acid in the body and can lead to RP, as well as other symptoms such as ataxia (difficulty with coordination), peripheral neuropathy, and cardiac issues.
* **Kearns-Sayre syndrome:** Kearns-Sayre syndrome is a rare mitochondrial disorder characterized by external ophthalmoplegia (paralysis of eye muscles), cardiac conduction defects, and pigmentary retinopathy resembling RP.
* **Abetalipoproteinemia (Bassen-Kornzweig disease):** This is an autosomal recessive disorder characterized by low or absent plasma cholesterol, low-density lipoproteins (LDL) and very low-density lipoproteins (VLDL). Other features include RP, acanthocytosis, spinocerebellar degeneration, and fat malabsorption.
* **Neurodegeneration with brain iron accumulation (NBIA):** Some forms of NBIA, a group of rare genetic disorders characterized by abnormal iron accumulation in the brain, may present with retinal abnormalities resembling RP. The most typical variant (pantothenate kinase-associated neurodegeneration, PKAN) shows the "eye of the tiger" sign on a T2-weighted magnetic resonance image due to a hyperintense signal at the center of the globus pallidus.
* Other systemic diseases associated with RP include familial isolated vitamin E deficiency and Alström syndrome.

These syndromes often have underlying genetic mutations that affect multiple organ systems, including the retina. It is important to note that the severity and specific clinical manifestations can vary widely among individuals with the same syndrome due to genetic heterogeneity and the influence of other genetic and environmental factors. Genetic testing and a multidisciplinary approach involving ophthalmologists, geneticists, and other specialists are typically necessary for accurate diagnosis and management of this condition.

**Epidemiology**

* Nonsyndromic RP has a worldwide prevalence of about 1 in 5000 individuals. RP constitutes around 50% of the cases of inherited retinal diseases, and affects more than 1.5 million people worldwide with varying prevalence. Estimates in prevalence range from 1 in 3026 in Denmark to 1 in 4869 in Birmingham, UK,and as high as 1 in 372 in rural India.Some of this variation may be due to differences in methodology and case definitions across studies; furthermore, the prevalence may be higher in some populations with more consanguineous marriages, as seen in certain Middle Eastern and South Asian countries.
* Men are affected slightly more often than women due to the X-linked form being expressed more frequently in males. Syndromic RP is much less common, with estimates for Usher syndrome ranging from 4 to 17 cases per 100,000 individuals.
* The average age of symptom onset is dependent on the genetic type involved. The autosomal recessive form will develop symptoms in the early adolescent years, but those affected with autosomal dominant RP will likely not have symptoms until well into their 20s. More than three-quarters of individuals with RP will be symptomatic and present for clinical evaluation and diagnosis of the disease by the time they are 30 years. In a study conducted in Japan, the average age of diagnosis was 35.1 years (median age 36.5 years).

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**SCLERITIS AND EPISCLERITIS**

**Definition and description**

Scleritis is the inflammation of your sclera, normally the white part of your eye. When you have scleritis, the white part of your eye becomes red.

Scleritis often involves piercing pain in your eye that gets worse with eye movement. It can cause permanent damage and vision loss. Treatment options include nonsteroidal anti-inflammatory drugs (NSAIDs) and corticosteroids. It may be caused by an underlying inflammatory disease, fungus or injury.

Scleritis should be treated. Don’t try to wait for it to go away on its own.

### **Types of scleritis**

There are two main types of scleritis: anterior (referring to the front of your sclera) and posterior (referring to the back of your sclera). Both anterior and posterior scleritis can also be diffuse, nodular or necrotizing.

* Diffuse scleritis: Diffuse scleritis is scattered all across your sclera. It’s the most common type.
* Nodular scleritis: Nodular scleritis is concentrated in one spot of the sclera. You can usually see the lump (nodule).
* Necrotizing scleritis: This most severe form of scleritis. It causes the most damage. It can destroy eye tissue and even result in the loss of your entire eye. There’s a form of necrotizing scleritis called scleromalacia perforans. It may not be painful but it can cause perforations (holes). This type accounts for 4% of scleritis.

Anterior scleritis, occurring at the front of the sclera, is the most common type of scleritis. Posterior scleritis, which affects the back of the sclera, represents about 10% of all cases of scleritis.

Scleritis usually affects people who are 47 to 60 years old. But people who are younger or older can get it, too. The condition is more common in females than in males, in part due to the association of scleritis with autoimmune conditions. But males tend to have higher rates of scleritis caused by an infection.

Each year, healthcare providers diagnose about 10,500 cases of scleritis in the United States. This works out to about 4-6 cases per 100,000 people.

### **What causes scleritis?**

Sometimes, scleritis has no known cause. Healthcare providers call this “idiopathic.” In many other cases, you may get scleritis if you have another type of medical condition, like an autoimmune illness. Other symptoms or conditions in this category include:

* Rheumatoid arthritis. This condition affects the joints on both sides of your body. Rheumatoid arthritis is the immune system condition most often associated with scleritis.
* Connective tissue diseases like systemic lupus erythematosus. Lupus and its chronic inflammation disrupt many parts of your body, which may include your joints, your skin and organs — like your lungs, brain, kidneys and heart.
* Inflammatory bowel disease (IBD). This term describes a group of disorders that cause chronic inflammation in your intestines.
* Sjögren’s syndrome. This autoimmune disorder restricts the amount of moisture provided by glands in your eyes and mouth.
* Scleroderma. In this disorder, normal tissue is replaced with dense, thick fibrous tissue.
* Granulomatosis with polyangiitis. This disease is the result of inflammation within your tissues (granulomatous inflammation) and blood vessels (vasculitis), which can damage organ systems.

Scleritis can also be associated with:

* Infections. Certain infections cause infectious scleritis. These can be bacterial, fungal or viral. Fungal infections usually have a less positive outlook than bacterial or viral infections. Lyme disease may also cause scleritis (Lyme scleritis).
* Trauma or injury to your eye, including surgical procedures. This type of injury often gives rise to infectious scleritis. In fact, one example of this is something called surgically induced necrotizing scleritis (SINS).
* Medications used to treat or prevent bone disease. These types of drugs include bisphosphonates and are known to cause inflammatory eye reactions.

**Signs and symptoms**

The signs and symptoms of scleritis include:

* Redness and swelling of your sclera.
* Pain and tenderness in your eye, often severe enough to wake you up at night. It can also spread to other parts of your face.
* Watering (tearing) eyes.
* Sensitivity to light (photophobia).

**Diagnosis methods**

Your provider may be able to determine if you have scleritis by giving you an eye exam, which may include a slit lamp exam.

If you have posterior scleritis, your provider may order a computed tomography (CT) scan or an ultrasound.  
If your scleritis is caused by an infection, your provider may take a smear of eye discharge to send to the lab. In very few cases, your provider might order a biopsy.

## **Management and Treatment**

If you have a very mild case of scleritis, your provider may recommend using nonsteroidal anti-inflammatory drugs (NSAIDS). But your provider is more likely to prescribe a systemic corticosteroid, like prednisone, for a longer period of time (seven to 10 days).

If you get inflammation again, you may need intravenous corticosteroids.

If you have an infectious version of scleritis, you may need an antibiotic, antifungal or antiviral.

For necrotizing scleritis, your provider may work with a rheumatologist to prescribe other medications, including immunotherapeutic drugs like cyclophosphamide, methotrexate, mycophenolate mofetil, or biologic agents like rituximab and adalimumab.

Other treatments may include scleral patching or grafts, which use other types of tissue as implants.

## **Prevention**

In many cases, you can’t prevent scleritis. But you can take good care of your eyes.

You can reduce your risk of damaging your eyes by wearing the required eye protection when you’re at work or participating in certain contact sports.

You can reduce your risk of eye infections by always making sure your hands are clean if you have to touch your eyes. Also, make sure you clean your contact lenses if you wear them.

## **Outlook / Prognosis**

Scleritis can — and should — be treated. Untreated scleritis can result in vision loss. Posterior necrotizing scleritis can be the most damaging. Your provider will also work with you to treat any other autoimmune conditions you may have.

## **Living With**

### **When should I see my healthcare provider about scleritis?**

You should see your healthcare provider any time you have pain, redness or swelling in one or both eyes. This is especially true if you have some type of immune system disorder.

**Differential diagnosis**

The most common differential diagnosis for patients with suspected infectious scleritis is autoimmune scleritis, as both manifest with similar signs and symptoms. Alternate diagnoses include episcleritis and anterior uveitis. Patients with systemic diseases such as vasculitis and syphilis have also been known to show symptoms similar to scleritis. A systemic workup can differentiate these from scleritis.

**Epidemiology**

One study reported that in 56 eyes with infectious scleritis, the median time from the inciting event was 1.9 months. Patients with a history of pterygium surgery had a median time of 49 months (range 0-183 months) after surgery before developing infectious scleritis. This duration is significantly longer than those with glaucoma, cataracts, and retinal surgery (median 1.0 to 1.6 months). The study also noted a median age of 70, suggesting that age might be a factor in developing this condition. The pathogens more commonly responsible for infectious scleritis are *Pseudomonas aeruginosa* in developed countries and *Nocardia* or fungi in developing countries.

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[Infectious Scleritis - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK560818/#article-109802.s4)

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### **Episcleritis**

Episcleritis is the medical name for inflammation (swelling), irritation and reddening of your episclera. Blood vessels in the eye get bigger, making it look red or pink. Episcleritis often affects only one eye but can affect both.

Your episclera is a layer of clear tissue that covers the white part of your eyes (sclera). It’s in between your sclera and the lining of your eyelids (the conjunctiva).

Episcleritis can affect anyone. But it usually happens more often in women between the ages of 47 and 60.

Each year in the United States, about 41 in 100,000 people are diagnosed with episcleritis.

### **Types of episcleritis**

There are two types of episcleritis. One is called simple episcleritis. You may have a limited red area in your eye (sectoral) or an area of redness may cover most of your eye (diffuse).

Episcleritis may start suddenly, which your healthcare provider will call “acute onset.” In simple episcleritis, your eye might get worse at 12 hours and then get better over two to three days.

Another type is nodular episcleritis. This type has a raised lump (nodule) of inflammation in your episclera. Nodular episcleritis often starts gradually rather than suddenly.

### **Signs and symptoms of episcleritis**

Signs and symptoms of episcleritis may include:

* Reddening and irritation of the whites of your eyes.
* Inflammation (swelling) of your eyes.
* Tearing (watering) of your eyes.
* Eye discomfort, but not actual pain.

### **Causes episcleritis**

In many cases, episcleritis has no known cause (it’s idiopathic). In other cases, episcleritis may be associated with inflammatory and immune system disorders. These types of diseases include:

* Rheumatoid arthritis. This is a type of arthritis where your immune system attacks the tissue lining your joints. Rheumatoid arthritis affects joints on both sides of your body.
* Lupus. Also known as systemic lupus erythematosus, lupus is an autoimmune disease that causes swelling and pain throughout your body. It can cause joint pain, skin issues and problems with organs.
* Inflammatory bowel disease (IBD). A group of disorders that cause pain and swelling in your intestines. If it lasts a long time (is chronic), it can damage the tissue.
* Rosacea. This condition primarily affects the skin on your face, causing redness that’s tough to get rid of. It can also cause problems with your eyes.
* Behçet’s disease. This is a chronic condition caused by inflammation of the blood vessels (vasculitis).

Some infections may cause episcleritis. These include:

* Lyme disease. This condition is caused by a bacterium (singular of bacteria). It’s transmitted by a tick bite.
* Syphilis. This is a sexually transmitted infection (STI) that can damage your health significantly.
* Herpes infections.
* Cat scratch fever, also called cat scratch disease. This infection is caused by a bacterium in cat saliva.

Sometimes healthcare providers note other factors — like stress, allergies and hormonal fluctuations — when a person has episcleritis. But they aren’t necessarily triggers of episcleritis.

## **Diagnosis and Tests**

Your eye care provider will most likely be able to diagnose episcleritis with an eye exam. They’ll also ask you about your health history, especially about any immune system disorders.

Your provider may need additional tests, like lab tests of your blood and/or imaging tests, to find out if you have an immune system or inflammatory disorder.

## **Management and Treatment**

Your provider may prescribe eye drops that contain corticosteroids. Your provider may also suggest using a nonsteroidal anti-inflammatory drugs (NSAIDs).

Using eye drops or NSAIDs can make the condition clear up more quickly.

If you have an issue with your immune system along with episcleritis, your provider will work with a rheumatologist to ensure that you get the treatment you need.

## **Prevention**

Since there’s often no way of telling what’s causing episcleritis, there’s no real way to prevent it from happening.

## **Outlook / Prognosis**

If you have episcleritis, the outlook is generally good. It’s not uncommon, though, for episcleritis to come back more than once.

If you have simple episcleritis, it’ll probably clear up on its own in two to three weeks. The eye drops or NSAIDs help your symptoms go away sooner.

There’ve been infrequent reports of complications from using steroids to treat episcleritis. These complications may include glaucoma or cataract development.

## **Living With**

### **How do I take care of myself if I have episcleritis?**

You can use cool compresses and chill your eye drops to make your eyes feel better.

If your provider agrees, you can take NSAIDs for the discomfort and inflammation.

### **When should I see my healthcare provider about episcleritis?**

You should see or talk to your healthcare provider at the beginning of an episcleritis episode, especially if you’ve never had this kind of thing happen before.

If you find that the efforts you’re making to manage episcleritis aren’t working, or if things get worse, you should see your healthcare provider immediately.

**Epidemiology**

Episcleritis is most commonly diagnosed in young to middle-aged females and is rarely diagnosed in children. There is no consensus on general population incidence and prevalence because these studies are not published in the literature. However, it is known that diffuse episcleritis is significantly more prevalent than the nodular form of the condition. Diffuse episcleritis occurs in about 70% of patients; whereas, nodular episcleritis occurs in only about 30% of patients.

It is also well established that the incidence and prevalence of episcleritis are higher in populations with systemic collagen-vascular disease and autoimmune diseases. In one study, recurrent episcleritis in the same or contralateral eye occurred in about 30% of patients. The global statistics may likely be more as patients may self-diagnose and treat recurrent episodes.

**DIFFERENTIAL DIAGNOSIS**

Misdiagnosis or delayed diagnosis of episcleritis is not common. The differential diagnosis list includes conditions that may appear to be similar to episcleritis, but after a thorough history and eye exam, misdiagnosis is uncommon.

Contact lens-associated red eye (CLARE) is a condition that may have a similar presentation to episcleritis but has a history and disease characteristics that make it difficult to mistake it for another condition. The patient affected by CLARE will have recently slept in their contact lenses and will have symptoms of unilateral pain, photophobia, and epiphora with normal visual acuity. The conjunctiva and cornea will show signs of inflammation, including corneal infiltrate and corneal edema and iritis, if severe.

A second condition on the differential diagnosis list should be acute conjunctivitis. Conjunctivitis is a broad term to include viral, bacterial, allergic/atopic, and toxic etiologies. Conjunctivitis most likely presents with an acute red eye with discharge, photophobia, itching/burning, and edematous eyelids. Depending on the specific cause, the patient will also exhibit conjunctival follicles, papillae, or a combination.

Sectoral conjunctival erythema and edema occur with phlyctenular conjunctivitis as well. This condition is caused by a delayed hypersensitivity reaction to antigens in the tear film and is often associated with blepharitis.

A mechanical inflammation of pinguecula causes pingueculitis. This is common when pingueculae are larger or co-occur with dry eye. Distinguishing features are that the inflamed area is associated with a pinguecula.

Iritis is another condition that may initially present similar to episcleritis, but the specific exam findings of iritis make it easy to differentiate. Patients with iritis will also have an acute onset of pain, redness, photophobia, and tearing. The conjunctival hyperemia is usually more concentrated in the circumlimbal area and is appropriately named ciliary flush. Distinguishing features of iritis are keratic precipitates on the corneal endothelium and cells and flare in the anterior chamber.

Scleritis is the most important condition to differentiate from episcleritis because the treatment of scleritis is more aggressive and can affect prognosis and complications. Patients with scleritis will complain of a more gradual onset of redness, pain, tearing, photophobia, and may have reduced visual acuity. These patients have deep, severe radiating pain from the affected eye, and their sclera will have a more red-purple hue. The hyperemia will not blanch with topical phenylephrine drops, and they may also have corneal involvement with peripheral stromal keratitis. More than 50% of patients with scleritis have a known systemic autoimmune connective tissue disease or vasculitis. The treatment regimen for scleritis includes topical steroids and oral NSAIDs similar to episcleritis, but patients often need to be treated with oral steroids or subconjunctival steroid injections. Severe cases may require management with immunosuppressants with potentially devastating associated side effects such as azathioprine, methotrexate, and mycophenolate mofetil.

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**STARGARDT DISEASE**

**Definition and description**

Stargardt disease is an eye condition that affects the macula and side parts of the retinas in your eyes. The macula is the part of your retina that lets you see objects directly in front of you (your central vision). Stargardt disease makes you lose your central vision over time. It can also affect your peripheral vision.

Stargardt disease is a type of macular degeneration. Most people with macular degeneration develop it later in life (after age 60). This is called age-related macular degeneration. Stargardt disease usually affects children and adults younger than 20. That’s why healthcare providers sometimes call it juvenile macular degeneration or juvenile macular dystrophy. It’s also known as fundus flavimaculatus when it affects the edges of your retina.

Stargardt disease happens when your body can’t get rid of a fatty yellow pigment called lipofuscin. If you have Stargardt disease, your body makes too much of it. The extra lipofuscin collects in your retina. It damages special cells called photoreceptors. Eventually, the damage permanently affects your vision.

### **Symptoms of Stargardt disease**

Stargardt disease will cause noticeable changes in your eyes and vision, including:

* Blind spots or cloudy areas in your vision
* Blurry vision
* Light sensitivity
* New color blindness
* Trouble adjusting to changes in lighting
* Worsened night vision

Stargardt disease symptoms are usually progressive. This means they get worse over time. Eventually, you’ll lose some or all of your central vision. You might still be able to see out of the sides of your eyes. But you won’t be able to clearly see objects directly in front of you.

### **Stargardt disease causes**

Stargardt disease is a genetic condition. It happens when there’s a genetic change in the *ABCA4* gene. This gene helps your retinas work correctly.

Stargardt disease is hereditary. Biological parents pass it to their children. Stargardt disease is passed through families in an autosomal recessive pattern. That means both biological parents need to have a changed *ABCA4* gene for their biological children to develop Stargardt disease.

## **Diagnosis and Tests**

### **How doctors diagnose Stargardt disease**

An eye doctor will diagnose Stargardt disease. They’ll give you an eye exam to check your vision and eye health. Tell your provider when you first noticed changes in your vision or other symptoms.

You might need a few tests, including:

* Electroretinography (ERG)
* Fluorescein angiography
* Fundus photography and fundus autofluorescent photography
* Genetic testing
* Optical coherence tomography (OCT)

#### **Stargardt disease stages**

Your eye care specialist may classify Stargardt disease with stages as it progresses, including:

* Stage 1. Flecks of excess lipofuscin form in your macula. You might have mild symptoms.
* Stage 2. The flecks have built up enough to spread beyond your macula to other areas of your retina around it. Symptoms will be more noticeable.
* Stage 3. The flecks have absorbed back into your macula and damaged it (they cause atrophy). This will cause worsening symptoms.
* Stage 4. The atrophy in your macula is severe enough to erase some or all of your central vision.

## **Management and Treatment**

There’s no treatment to reverse Stargardt disease or the damage it causes. Your eye care specialist will suggest ways to manage symptoms and slow down the vision loss, including:

* Avoiding dietary supplements that contain vitamin A (getting too much vitamin A may speed up vision loss if you have Stargardt disease)
* Glasses, contacts and/or specific low vision aids
* Quitting smoking (and other forms of nicotine, like vaping)
* Wearing a hat or sunglasses to protect your eyes when you go outside

#### **Is Stargardt disease curable?**

Your eye care specialist can help you manage Stargardt disease, but you can’t cure it.

Researchers are conducting clinical trials to find ways to treat all types of macular degeneration (including Stargardt disease). Ask your eye care specialist if you’d be a good fit for a clinical trial. This can help you get access to new, experimental treatments.

## **Outlook / Prognosis**

You should expect to lose your central vision. Most people experience vision loss slowly over several years. It sometimes takes decades. But it can happen faster. How your *ABCA4* gene is changed can affect how quickly you lose your vision. This can also control how much of your vision you lose. Your eye doctor will help you understand what to expect.

You’ll need regular eye exams to monitor changes in your eyes. Ask your eye care specialist how often you should schedule routine exams and any follow-up tests.

## **Living With**

### **How do I take care of myself with Stargardt disease?**

You can take care of yourself in many ways by eating a nutritious diet and getting enough exercise. Don’t smoke. Deal with stress as it comes. Keep your regular schedule of appointments with your healthcare providers.

### **When should I see my healthcare provider if I have Stargardt disease?**

Your eye care specialists will want to see you on a regular basis. Make sure you keep those appointments. However, if you have any changes in vision or any type of pain, you should contact your provider right away.

**Differential diagnosis**

Several inherited macular dystrophies have a clinical appearance similar to STGD1. Distinguishing STGD1 among these similar-appearing conditions is essential for prognostication, systemic associations, and genetic counseling.

**Pattern Dystrophy**

Marmor and Byers were the first to propose “pattern dystrophy” when describing a spectrum of characteristic fundus patterns (ie, butterfly, reticular pigmentary). Pattern dystrophy (PD) most commonly masquerades as STGD1. PD is an autosomal dominant condition with mild or no visual loss in the second to fifth decade. PD associated with the *PRPH2* gene (multifocal pattern dystrophy simulating fundus flavimaculatus) primarily manifests as a macular disease. However, the phenotype may range from macular flecks to diffuse pigmentation in the periphery. Peripapillary sparing is a characterization of PD similar to STGD1. However, PD is associated with a higher incidence of CNVM. FFA helps distinguish the two as there will be window defects in PD, in contrast to a “masked choroid” in STGD1.

**Autosomal Dominant Stargardt-like Macular Dystrophies**

These dystrophies include variants associated with the *ELOVL4* (STGD2 or STGD3) and the *PROM1* (STGD4) genes.

**Batten Disease/Juvenile Neuronal Ceroid Lipofuscinosis**

Defects in the processing of lysosomal storage characterize Batten disease/juvenile neuronal ceroid lipofuscinosis, a rare metabolic disease. These defects lead to apoptosis of both the photoreceptor (retina) and the neuronal (brain) cells. Visual loss and severe color vision deficiency are common, and the life expectancy of patients with this condition is usually 20 to 30 years.

The disease progression is usually rapid. Ocular examination shows vascular attenuation and optic disc pallor. OCT reveals a thinned retinal nerve fiber layer, while dark-adapted full-field ERG shows an absent or electronegative wave. Maintain high suspicion in younger patients diagnosed with severe STGD1 with rapid progression.

**Age-Related Macular Degeneration**

This condition can mimic late-onset STGD1, as the flecks may be mistaken for drusens. A careful examination can help distinguish the two, as drusen tend to become confluent toward the center of the macula. Flecks show intense hyper AF, while drusens may be mildly hyper autofluorescence or hypo autofluorescence. Sub-RPE accumulation on OCT characterizes drusens, while STGD1 is associated with hyperreflective thickening of the RPE. Age-related macular degeneration does not show a dark choroid on FFA.

**Pentosan Polysulfate Maculopathy**

This condition is associated with the intake of pentosan, used in treating interstitial cystitis. The FAF changes may not spare the peripapillary region in pentosan polysulfate maculopathy

### **Epidemiology**

Although Stugardt disease is the most common cause of juvenile macular dystrophy, its prevalence in the United States is estimated to be 10 to 12.5 per 100,000 individuals. Cornish et al.estimated the annual incidence of this condition in the United Kingdom to be around 0.110 to 0.128 per 100,000 individuals. A study conducted in the Netherlands found the incidence of Stargardt disease to be 1.67 to 1.95 per 1,000,000 individuals per year. The point prevalence of Stargardt disease was 1:22,000 to 1:19,000 in 2018 in the Netherlands

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**STRABISMUS**

*ALTERNATIVE NAMES: Strabismus is also known as heterotopia, squint, cast, cross-eyes, wall-eyes, esotropia, exotropia, hypotropia, hypertropia, misalignment of the eyes.*

**DEFINITION / DESCRIPTION**

This is a disorder in which there is a misalignment or deviation of the eyes with each other, and they do not line up in the same direction, they do not look at the same object at the same time, which can lead to double vision or amblyopia(lazy eye) if untreated. The most common form of strabismus is known as “crossed eyes”

Other disorders associated with strabismus in children include:

* Apert syndrome
* Cerebral palsy
* Congenital rubella
* Hemangioma near the eye during infancy
* Incontinentia pigmenti syndrome
* Noonan syndrome
* Prader-Willi syndrome
* Retinopathy of prematurity
* Retinoblastoma
* Traumatic brain injury
* Trisomy 18

**CAUSES**

Six different muscles surround each eye and work "as a team." This allows both eyes to focus on the same object.

In someone with strabismus, these muscles do not work together. As a result, one eye looks at one object, while the other eye turns in a different direction and looks at another object.

When this occurs, two different images are sent to the brain -- one from each eye. This confuses the brain. In children, the brain may learn to ignore (suppress) the image from the weaker eye.

If the strabismus is not treated, the eye that the brain ignores will never see well. This loss of vision is called amblyopia. Another name for amblyopia is "lazy eye." Sometimes the lazy eye is present first, and it causes strabismus.

In most children with strabismus, the cause is unknown. In more than one half of these cases, the problem is present at or shortly after birth. This is called congenital strabismus.

Most of the time, the problem has to do with muscle control, and not with muscle strength.

Strabismus that develops in adults can be caused by:

* Botulism
* Diabetes (causes a condition known as acquired paralytic strabismus)
* Graves disease
* Guillain-Barré syndrome
* Injury to the eye
* Shellfish poisoning
* Stroke
* Traumatic brain injury
* Vision loss from any eye disease or injury

**RISK FACTORS**

A family history of strabismus is a risk factor. Farsightedness may be a contributing factor, often in children. Any other disease that causes vision loss may also cause strabismus.

Strabismus is more prevalent with certain syndromes like Down syndrome, cerebral palsy, Apert-Crouzon syndrome, premature infants with low birth weight, and children with affected parents or siblings. All siblings of a strabismic child should be screened at an early age for strabismus as sensorimotor anomalies are common in the pedigrees of strabismic probands.

**SIGNS / SYMPTOMS**

Symptoms of strabismus may be present all the time or may come and go. Symptoms can include:

* Crossed eyes
* Double vision
* Eyes that do not aim in the same direction
* Uncoordinated eye movements (eyes do not move together)
* Loss of vision or depth perception

It is important to note that children may never be aware of double vision. This is because amblyopia can develop quickly.

**DIAGNOSIS METHOD (EXAMS AND TESTS)**

The health care provider will do a [physical exam](https://medlineplus.gov/ency/article/002274.htm). This exam includes a detailed examination of the eyes.

The following tests will be done to determine how much the eyes are out of alignment.

* Corneal light reflex
* Cover/uncover test
* Retinal exam
* Standard ophthalmic exam
* Visual acuity

A brain and nervous system (neurological) exam will also be done.

**TREATMENT OPTIONS**

The first step in treating strabismus in children is to prescribe glasses, if needed. Next, amblyopia or lazy eye must be treated. A patch is placed over the better eye. This forces the brain to use the weaker eye and it develops better vision.

Your child may not like wearing a patch or eyeglasses. A patch forces your child to see through the weaker eye at first. However, it is very important to use the patch or eyeglasses as directed.

Eye muscle surgery may be needed if the eyes still do not move correctly. Different muscles in the eye will be made stronger or weaker. To strengthen a muscle, it is removed from the eye, shortened, then reattached. To weaken a muscle, it is removed from the eye and reattached further toward the back of the eye. Often in adults, an adjustable suture method is used so that the final adjustment of the position of the weakened muscle is made with the person awake and looking at a target. This has been shown to be more accurate.

Eye muscle repair surgery does not fix the poor vision of a lazy eye. [Muscle surgery](https://medlineplus.gov/ency/article/002961.htm) will fail to improve vision if amblyopia has not been treated. A child may still have to wear glasses after surgery. Surgery is more often successful if done when the child is younger.

Adults with mild strabismus that comes and goes may do well with glasses. Eye muscle exercises may help keep the eyes straight. More severe forms will require surgery to straighten the eyes. If strabismus has occurred because of vision loss, the vision loss will need to be corrected before strabismus surgery can be successful.

**Adult Strabismus (Crossed Eyes) Treatment**

There are several ways to treat strabismus in adults; they may include:

**Adult strabismus (crossed eyes) surgery**

This is the most common treatment for strabismus. Surgery can improve eye alignment and help restore proper vision.

Typically, strabismus occurs when the [muscles around the eyes](https://www.aao.org/eye-health/anatomy/eye-muscles) are either too stiff or too weak. An ophthalmologist can loosen, tighten, or move certain eye muscles so that the eyes line up properly to work together. **More than one surgery may be needed to treat strabismus**.

Surgery is usually done as outpatient surgery in a hospital or surgery center, using either general or local anesthesia. Your ophthalmologist makes a small cut in the tissue covering the eye to reach the eye muscles. The muscles are then repositioned to help the eyes point in the same direction. This may need to be done in one or both eyes. After strabismus surgery, you can get back to your daily routine within a few days.

**Eye muscle exercises**

An ophthalmologist can teach you exercises to help you focus both eyes inward. These exercises can help if you have “[convergence insufficiency](https://www.aao.org/eye-health/diseases/what-is-convergence-insufficiency).” That is when your eyes do not align properly for close tasks, like reading or computer work.

**Prism eyeglasses**

A prism is a clear, wedge-shaped lens that bends (refracts) light rays. A prism can be attached to eyeglasses or made as part of the lens. Prisms can help some people with mild double vision see one image, not two.

**Botulinum toxin (Botox®)**

In some cases, an injection (shot) of this drug in the eye muscles can help treat strabismus. It paralyzes the muscles that keep your eyes from aligning properly. The effect can last for just a few months, or it could permanently improve eye alignment.

**OUTLOOK / PROGNOSIS**

After surgery, the eyes may look straight, but vision problems can remain.

The child may still have reading problems in school. Adults may have a hard time driving. Impaired vision may affect the ability to play sports.

In most cases, the problem can be corrected if identified and treated early. Permanent [vision loss](https://medlineplus.gov/ency/article/003029.htm) in one eye may occur if treatment is delayed. If amblyopia is not treated by about age 11 years, it is likely to become permanent, However, new research suggests that a special form of patching and certain medicines may help to improve amblyopia, even in adults. About one third of children with strabismus will develop amblyopia.

Many children will get strabismus or amblyopia again after successful treatment. Therefore, the child will need to be monitored closely.

**WHEN TO SEE A DOCTOR/MEDICAL PROFESSIONAL**

Strabismus should be evaluated promptly. Contact your provider or eye doctor if your child:

* Appears to be cross-eyed
* Complains of double vision
* Has difficulty seeing

**Note**: Learning and school problems can sometimes be due to a child’s inactivity to see the blackboard or reading material.

**There Are Six Eye Muscles That Control Eye Movement**

One muscle moves the eye to the right, and one muscle moves the eye to the left. The other four muscles move the eye up, down, and at an angle. In order to focus on a single image, the muscles from both eyes must work together.

**How Does Adult Strabismus Affect Vision?**

With normal vision, both eyes aim at the same spot. The brain combines the two images from our eyes into a single, three-dimensional (3-D) image. This is how we can tell how near or far something is from us (called depth perception).

When one eye is out of alignment, two different pictures are sent to the brain. In a young child, the brain learns to ignore the image of the misaligned eye. Instead, it sees only the image from the straight or better-seeing eye. As a result, the child loses depth perception.

Adults who develop strabismus after childhood often have double vision. This is because their brains have already learned to receive images from both eyes. Their brains cannot ignore the image from the turned eye, so they see two images.

**DIFFERENTIAL DIAGNOSIS**

Differential diagnoses can be classified based on the type of defect. Important differentials to be considered are as follows:

**Congenital Esotropia**

* Early-onset accommodative esotropia
* Abducens palsy
* Nystagmus blockage syndrome
* Duane retraction syndrome
* Sensory esotropia
* Strabismus fixus
* Moebius syndrome

**Fully Accommodative Esotropia**

* Non-accommodative esotropia
* Congenital esotropia
* Cyclic esotropia
* Convergence excess and near esotropia

**Intermittent Exotropia**

* Infantile exotropia
* Convergence weakness or insufficiency
* Sensory exotropia with poor unilateral vision

**It Is Never Too Late to Treat Strabismus**

You do not have to live with the discomfort and problems caused by misaligned eyes. With your ophthalmologist’s help, you can find a treatment for your strabismus.

**EPIDEMIOLOGY**

The prevalence of strabismus is 2% to 5% in the general population.. In the U.S., 5 to 15 million individuals are affected by strabismus. In a National Health Survey, exotropia was seen in 2.1% and esotropia in 1.2% of the population aged 4 to 74 years. This difference is due to the higher prevalence of exotropia in the population between 55 to 75 years of age.

Fifty percent of all childhood esotropias are either fully or partially accommodative. Non-accommodative esotropia is seen in 10% of all strabismus cases and is the second most common form of childhood esotropia. Infantile esotropia affects 1 in every 100 to 500 persons, which accounts for 8.1% of cases of esotropia.

Intermittent esotropia is seen in 1% of the population and is the most common form of exotropia. Exotropia is more prevalent in Asian and black populations. Women make up 60% to 70% of exotropia cases.

**REFERENCES:** [Strabismus - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK560782/)

**STYE (HORDEOLUM)**

**Definition and description**

A stye (or sty) is a painful red bump on the edge of your eyelid. It can look similar to an acne pimple and may be tender to the touch. A stye forms when a tiny oil-producing gland in your eyelash follicle or eyelid skin becomes blocked and a bacterial infection develops. The medical term for a stye is “hordeolum.”

It’s common to have a stye on only one eyelid, but it’s also possible to get styes on both lids. A stye usually lasts one to two weeks and will typically go away on its own. But in cases where it doesn’t, you may need to rely on an eye care provider to drain it. They may also prescribe antibiotics to reduce the infection.

A stye is similar to another eyelid bump called a chalazion. A chalazion is a bump that usually occurs farther back on your eyelid. Unlike a stye, a chalazion usually isn’t painful and isn’t caused by a bacterial infection. But treatment for both conditions is similar.

**Types of styes**

There are two types of styes:

External styes: External styes form on the outer part of either your upper or lower eyelid. They’re the more common type. An infection in your eyelash follicle usually causes them.

Internal styes: Internal styes form on either of your inner eyelids (facing your eyeball). An infection in the inner eyelid gland that produces oils that help keep your eyelid moist causes this type.

Styes are very common. They’re more common in adults than children because the oil in an adult’s oil glands is thicker than a child’s. That means it’s more prone to blockage.

**What causes a stye?**

A bacterial infection in your eyelid’s oil-producing glands causes most styes. The oil-producing glands line the eyelids and help lubricate the surface of your eye.

Are styes contagious?

Styes generally aren’t contagious. But small amounts of bacteria can be spread from them. This is why it’s important to always wash your hands before and after touching a stye and wash pillowcases often to help prevent the bacteria from spreading. Unless you’re cleaning or applying warm compresses to the stye, avoid touching it to reduce bacteria spread and irritation.

**What are the risk factors for developing a stye?**

Styes are very common, and anyone can get them. But you may be more likely to get a stye if you:

Have had a stye before.

Have blepharitis (an inflammation of your eyelids).

Have certain skin conditions, like acne, rosacea or dandruff (seborrheic dermatitis).

Have diabetes.

Have dry skin.

Are experiencing hormonal changes.

Have high lipid levels (“bad” cholesterol).

**Signs and symptoms**

Is That Bump on Your Eye a Stye or a Chalazion?

What are the symptoms of a stye?

The main symptom of a stye is a painful red bump along your eyelid edge near your eyelashes. Other stye symptoms may include:

Swelling of your eyelid (sometimes your entire eyelid).

Discharge from your eye.

Crusting along your eyelid.

Light sensitivity.

Soreness and itching.

Eye tearing.

A scratchy feeling or a feeling that there’s something in your eye.

**Diagnosis methods (tests, lab work, imaging, etc.)**

Some styes are more stubborn and require a visit to a healthcare provider. If your vision seems to be affected or if your stye seems to be getting worse instead of better, contact a provider.

During your appointment, your provider will examine your eyelid and ask about any additional symptoms you’re having. They’ll be able to diagnose a stye based on this eye exam.

**Management and Treatment**

How do you get rid of a stye?

A stye will usually go away by itself in one to two weeks. To feel better faster and reduce pain and swelling, you can use a self-care plan to treat your stye at home. Here are some dos and don’ts to manage your stye at home.

**Do**

Use warm compresses. Apply a warm washcloth to your eyelid for 10 to 15 minutes at a time, three to five times a day. Rewarm the washcloth by soaking it in warm water, wring and repeat. Many people believe that using green tea bags moistened in warm water as eye compresses will help the stye not only feel better, but also speed healing, due in part to the antibacterial properties of green tea. Some scientists have shown that a natural antioxidant in green tea breaks down the cell wall of the bacteria, killing it. While there’s debate about this among eye experts, it won’t hurt you and should be at least as effective as using a warm washcloth as a compress.

Clean eyelids. Gently wipe away eye discharge with a mild soapy solution made from half baby shampoo and half water. You can also use eyelid wipes available in most drug stores.

**Don’t**

Squeeze or pop a stye.

Rub or touch your eyelid.

Wear makeup or contact lenses until the area has healed.

How will an eye care provider treat a stye?

If after 48 hours of stye self-care, your pain and swelling aren’t getting any better, it’s time to call your eye care provider. Stye treatment by a medical provider may include:

A small cut (incision) to drain your stye in the office (under local anesthesia).

Prescription antibiotic ointment to apply to your eyelid or antibiotic eye drops. Your provider may prescribe oral antibiotics in cases where the area around your eye is infected or after an incision is made to drain an internal stye.

A steroid injection into the stye to reduce eyelid swelling.

**Prevention**

The best way to prevent a stye is to practice good facial hygiene, including:

Wash your hands thoroughly and often, especially before touching your face and eyes.

Wash your hands before and after removing contact lenses. Clean your contacts with disinfectant and lens cleaning solution. Dispose of daily wear or other “limited use” lenses on the schedule that your eye care provider recommends.

Washing your face to remove dirt and/or makeup before going to bed.

Throwing away eye makeup every two to three months. Never share eye makeup with anyone else.

**Outlook / Prognosis**

How serious is a stye?

Styes are usually harmless. They may cause some minor irritation and discomfort, but they typically go away on their own. Stye self-care measures like warm compresses can help speed up the healing process.

Although it will be tempting to cover the stye with makeup, avoid doing this. Putting makeup on a stye can delay the healing process or even cause it to become more plugged up and infected, which, in turn, will make it more painful.

**Living With**

When should I see my healthcare provider?

You should see your healthcare provider if:

Your eye is swollen shut.

Pus or blood is leaking from the bump.

Pain and/or swelling increases after the first two to three days.

Blisters have formed on your eyelid.

Your eyelids feel hot.

Your vision has changed.

Styes keep coming back. If this happens, your provider may take a biopsy (a small sample of the stye), under local anesthesia, to rule out other more serious problems.

**DIFFERENTIAL DIAGNOSIS**

A stye presents as a localized, painful, red, and swollen lesion on the eyelid. Conditions that can mimic this clinical entity must be ruled out to ensure accurate diagnosis and management. Below is a list of key differential diagnoses.

* Chalazion: A chalazion is a chronic, nontender, firm nodule resulting from the blockage of a meibomian gland. Chalazia are typically painless and do not present with acute inflammation, which distinguishes them from styes, which are acute. Chalazia are chronic and often follow the resolution of a hordeolum.
* Preseptal cellulitis: Preseptal cellulitis is a bacterial infection of the eyelid and periorbital tissues anterior to the orbital septum. This condition is characterized by diffuse eyelid swelling, redness, and tenderness but lacks a localized nodule or abscess. Unlike chalazia, preseptal cellulitis does not present with a fluctuant nodule or pus discharge and may be accompanied by systemic symptoms such as a low-grade fever.
* Orbital cellulitis: This severe bacterial infection affects the tissues posterior to the orbital septum. Orbital cellulitis is distinguished by significant proptosis, restricted extraocular movement, and vision changes. Systemic symptoms such as high fever and malaise often accompany this pathology, making immediate imaging and hospitalization essential.
* Sebaceous gland carcinoma: This rare but serious malignant tumor of the sebaceous glands often mimics a recurrent chalazion or hordeolum. Key differentiating features include a persistent or recurrent lesion that does not respond to appropriate treatment and the presence of eyelid margin abnormalities such as loss of eyelashes (madarosis). A biopsy is required for definitive diagnosis.
* Blepharitis: This condition involves inflammation of the eyelid margin, often due to bacterial infection or meibomian gland dysfunction. Blepharitis is characterized by diffuse redness and scaling along the lid margins, often involving both eyes. This chronic condition is marked by irritation and crusting rather than acute painful swelling.
* Dermoid cyst: This congenital lesion is commonly found along the orbital rim. A dermoid cyst is distinguished by being painless and noninflammatory, presenting as a firm and well-circumscribed mass.
* Dacryocystitis: This condition involves infection of the lacrimal sac following obstruction of the nasolacrimal duct. Dacryocystitis presents with swelling, redness, and tenderness localized to the inner corner of the eye near the lacrimal sac, often accompanied by tearing or purulent discharge.
* Herpes simplex and zoster ophthalmicus: Herpes simplex is caused by the herpes simplex virus, while herpes zoster ophthalmicus arises from the reactivation of the varicella-zoster virus. These viral infections lead to inflammation of the eyelid. Herpes simplex is characterized by vesicles on the eyelid, while herpes zoster presents as a vesicular rash along the trigeminal nerve distribution, often accompanied by crusting and pain.
* Molluscum contagiosum: This viral infection is caused by the molluscum contagiosum virus, presenting as dome-shaped, pearly papules on the eyelid. The lesions are painless and feature a central umbilication, distinguishing them from other conditions. Unlike styes and chalazia, molluscum contagiosum lesions do not show signs of inflammation or acute tenderness.

Accurate identification of the condition is essential to avoid inappropriate management, particularly with serious conditions like orbital cellulitis and sebaceous gland carcinoma, which require urgent treatment. When in doubt, referral to an ophthalmologist and additional investigations such as imaging or biopsy may be required. This structured approach ensures proper diagnosis and treatment while minimizing the risks of mismanagement.

**EPIDEMIOLOGY**

While hordeola are very common, the exact incidence is unknown. Every age and demographic is affected, although a slightly increased incidence is observed among patients aged 30 to 50. Prevalence differences among populations worldwide are unknown. Patients with chronic conditions such as seborrhoeic dermatitis, diabetes, and hyperlipidemia may also be at increased risk.

**Prevalence in Different Regions**

Styes are common worldwide, affecting individuals of all ages. While the exact prevalence is not well-documented, the occurrence is significantly influenced by risk factors such as poor eyelid hygiene, blepharitis, and meibomian gland dysfunction. Styes are more frequently observed in regions with poor access to healthcare or suboptimal hygiene practices.

**Sex Distribution**

Styes are slightly more common in female individuals. This trend is attributed to the frequent use of cosmetics and eye makeup in this group, which can block gland ducts and exacerbate bacterial colonization.

**Age Distribution**

Styes can occur in all age groups, but certain factors increase the risk for some individuals. Frequent eye rubbing and inadequate eyelid hygiene can make children and adolescents vulnerable to developing a stye. Conditions that increase susceptibility to hordeolum formation in adults include chronic blepharitis, rosacea, and meibomian gland dysfunction. Age-related changes in meibomian gland function and tear production can cause recurrent styes in older adults.

**Associated Risk Factors**

Styes are more frequent in individuals with the following conditions:

* Blepharitis, which causes chronic eyelid inflammation
* Diabetes, which impairs the immune response
* Rosacea, which affects meibomian gland function and increases susceptibility to eyelid infections

By understanding the epidemiology of styles, healthcare providers can better identify at-risk populations and emphasize preventive strategies, including proper eyelid hygiene and timely treatment of underlying conditions.

REFERENCES

[Hordeolum (Stye) - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/sites/books/NBK459349/#article-29591.s4)

**THYROID EYE DISEASE**

**Definition and description**

Thyroid eye disease also is called Graves' ophthalmopathy. About 25% of people with Graves' disease have eye symptoms. Thyroid eye disease affects muscles and other tissues around the eyes. Symptoms may include:

* Bulging eyes.
* A gritty feeling in the eyes.
* Pressure or pain in the eyes.
* Puffy eyelids or eyelids that don't cover the eyeball all the way. This is called retracted eyelids.
* Red or inflamed eyes.
* Light sensitivity.
* Blurred or double vision.
* Vision loss.

Thyroid eye disease (TED) is an inflammatory disorder that affects the tissues around your eyes, causing swelling, discomfort and other symptoms. It happens in some people who have autoimmune diseases that attack their thyroid gland. Most often, it happens in people who have Graves’ disease. In this case, your healthcare provider may call it Graves’ eye disease.

Although autoimmune diseases are lifelong, TED has distinct phases. It may be active for two years or more before the inflammation subsides. During this time, symptoms may come and go and may be mild to severe. TED is mild for most people. But when it’s severe, it can cause permanent damage, with lasting cosmetic and vision changes.

Ophthalmopathy and orbitopathy are two other terms for eye disease. Your healthcare provider may describe TED as:

* Thyroid ophthalmopathy
* Thyroid orbitopathy
* Graves’ ophthalmopathy
* Graves’ orbitopathy

### **Causes of thyroid eye disease**

Thyroid eye disease is an autoimmune disease. This means your immune system attacks your thyroid and eye tissues. Graves’ disease is a common cause, but other autoimmune thyroid diseases, like Hashimoto’s disease, can also cause it. Even people with normal thyroid function can develop TED.

In these diseases, your immune system creates antibodies that mimic thyroid hormones and attach to thyroid hormone receptors. These receptors are mostly within your thyroid itself, but some are in the tissues behind your eyes. So, the same antibodies that affect your thyroid can also affect your eyes.

#### **Thyroid eye disease risk factors**

You may be more likely to get thyroid eye disease (Graves’ eye disease) if you:

* Are female. Females are five times more likely to get Graves’ disease and Graves’ eye disease. (But when males get it, it’s more likely to be severe.)
* Have high or low thyroid hormone levels. Abnormal thyroid hormone levels can contribute to TED. These hormones stimulate the same receptors in your eye tissues that the antibodies do.
* Have had radioiodine therapy. RAI therapy is a standard treatment for overactive thyroid (hyperthyroidism), which Graves’ disease causes. But it may contribute to TED or make it worse.
* Smoke. Smoking and secondhand smoke exposure significantly raise your risk of thyroid eye disease. Smoke exposure also makes you more likely to have more severe symptoms that last longer. If you have TED, the best thing you can do for your disease is quit smoking.

### **Thyroid eye disease symptoms**

Typical thyroid ophthalmopathy symptoms include:

* Bulging eyes (proptosis)
* Eye irritation
* Swollen and inflamed eyelids (blepharitis)
* Dry eyes or teary eyes
* Frequent blinking
* Light sensitivity (photophobia)
* Eye pain and headaches
* Difficulty moving your eyes
* Double vision (diplopia)

Symptoms normally affect both eyes, but sometimes, you may only notice symptoms in one eye.

## **Diagnosis and Tests**

### **What tests will be done to diagnose thyroid eye disease?**

A healthcare provider will be able to diagnose thyroid eye disease by doing a physical eye exam. They’ll be able to examine both your eyelids and your eyes.

If your healthcare provider thinks that you have thyroid eye disease, they’ll order blood tests to check if your thyroid hormone levels and antibodies are too high or too low.

Other tests your provider may request include:

* Ultrasound of the eyes
* Computed tomography (CT)
* Magnetic resonance imaging (MRI)

## **Management and Treatment**

### **What is the treatment for thyroid eye disease?**

The treatment you’ll need will depend on what stage the disease is in and how severe it is. Supportive treatments can help ease your symptoms while thyroid eye disease is active. These might include home remedies, over-the-counter or prescription medications. After the active phase has ended, you might need or want cosmetic surgery or vision correction surgery.

Supportive treatments for thyroid eye disease may include:

* Eye drops. During the active phase of TED, eye drops can help relieve your symptoms. Different types of eye drops can help with different symptoms, like dryness, redness or pain.
* Selenium supplements. Selenium deficiency increases TED symptoms.
* Scleral lenses. Similar to contact lenses, these lenses cover more of the surface of your eye, offering protection from the elements. You can also use them to apply medication.
* Vision aids. If you have double vision, your eye care provider may suggest wearing an eye patch or special eyeglasses with prisms to help correct it while thyroid eye disease is active.
* Thionamides. If you have hyperthyroidism (Graves’ eye disease), you may need medications to reduce your thyroid hormone levels. Examples include methimazole and propylthiouracil.
* Corticosteroids. When TED symptoms are more severe, healthcare providers prescribe a short course of corticosteroids to bring the inflammation down. You may take them by mouth or IV (intravenously, through a vein).
* Teprotumumab (Tepezza®). This newer drug became the first medication specifically approved to treat thyroid eye disease in the U.S. in 2020. It’s an alternative anti-inflammatory drug within the biologics category. More drugs like it are in development.
* Radiation therapy. Your eye doctor may recommend that you see a radiation oncologist to consider radiation, which reduces inflammation by killing the immune cells that are active behind your eyes.

Your healthcare team can offer alternative treatments if these don’t work for you. Rarely, some people may need surgery during the active phase of Graves’ eye disease. But in most cases, you and your team will consider surgery after your condition has stabilized. At this point, it’ll be easier to tell which appearance and vision changes are likely to be permanent.

#### **Surgery for Graves’ eye disease**

Surgery for thyroid eye disease may include:

* Orbital decompression surgery. This eye surgery may be necessary in the rare event that inflammation compresses your optic nerve. A surgeon will remove bone to reduce the compression. This surgery may also be done to reduce eye bulging after your condition has stabilized.
* Thyroidectomy. This treats severe hyperthyroidism. If your healthcare team can’t control your thyroid hormone levels with medications, they might need to remove your thyroid gland.
* Eyelid surgery. If your eyelids have retracted, you might need eyelid surgery to reposition them. This can improve your comfort as well as appearance and protect your eyes from exposure.
* Eye muscle surgery. You might need surgery on your eye muscles if they’re scarred and restricting your eye movement, or you have double vision because your eyes are misaligned.
* Oculoplastic surgery. Oculoplastic surgery adjusts the soft tissues and/or bones around your eyes. It can address cosmetic concerns like bulging eyes and bags around your eyes.
* Corneal transplant. If your cornea was badly damaged by exposure during the active phase of Graves’ eye disease, you might consider replacing it with a transplant to improve your vision.

## **Outlook / Prognosis**

### **What is the outlook for thyroid eye disease?**

The outlook is good for most people with thyroid eye disease. Most have mild symptoms, which often resolve on their own. People over the age of 50 tend to have more severe symptoms. If you have a serious case of TED, you might need a combination of treatments to manage it. Some people have lasting changes to their eyes, which may require surgery.

Prevention tips

### **Complications of Graves’ eye disease**

Appearance and vision changes that occur with thyroid eye disease may improve after the active phase is over, but they don’t always. Scarring can prevent the tissues around your eyes from healing and returning to their normal shape. Sometimes, parts of your eyes that affect your vision are permanently damaged. Surgery can treat some of these effects.

Lasting appearance changes may include:

* Eyelid retraction
* Protruding eyes
* Baggy eyes
* Red eyes

Lasting vision changes may include:

* Blurry vision
* Double vision
* Severe vision loss

When to see a doctor / red flag

#### **When should I call my healthcare provider?**

When you have thyroid eye disease, it’s important to let your healthcare team know if your symptoms worsen. You should also let them know if you notice any changes in your vision. Some changes might need urgent treatment. Call right away if you notice:

* Your field of vision has narrowed or closed off
* Colors appear differently than they used to
* Sudden, severe eye pain

**Differential diagnosis**

## Orbital Tumors

* Usually cause unilateral proptosis, whereas TED is typically bilateral (though it can be asymmetric or unilateral in some cases).
* May present with pain, vision loss, or rapidly progressive proptosis.
* Imaging (CT/MRI) shows mass lesions distinct from the muscle enlargement pattern seen in TED.

## 2. Idiopathic Orbital Inflammatory Syndrome (Orbital Pseudotumor)

* Presents with painful orbital inflammation, swelling, and restricted eye movements.
* Unlike TED, it often involves the tendinous insertions of extraocular muscles (TED typically spares tendons).
* Rapid onset and response to steroids are typical.
* May be unilateral or bilateral.

## 3. Orbital Myositis

* Inflammatory enlargement of one or more extraocular muscles causing pain, diplopia, and swelling.
* Usually unilateral and painful.
* Differentiated by imaging showing muscle belly and tendon involvement.

## 4. Carotid-Cavernous Fistula

* Presents with red eye, pulsatile proptosis, conjunctival chemosis, and bruit.
* Often unilateral.
* Associated with increased venous pressure rather than autoimmune inflammation.

## 5. Lymphoproliferative Disorders (Orbital Lymphoma)

* Usually painless, slowly progressive proptosis.
* Can mimic TED but typically lacks inflammatory signs like eyelid retraction or conjunctival injection.
* Imaging shows well-defined orbital masses.

## 6. Allergic Conjunctivitis

* May cause periorbital swelling and conjunctival injection.
* Symptoms include itching, which is uncommon in TED.
* No proptosis or lid retraction.

## 7. Dry Eye Syndrome

* Common in TED but also a separate diagnosis.
* Presents with irritation, redness, and grittiness without proptosis or eyelid retraction.

## 8. Chronic Progressive External Ophthalmoplegia

* Characterized by slowly progressive bilateral ptosis and ophthalmoplegia.
* No proptosis or inflammatory signs.

## 9. Obesity and Other Causes of Pseudoproptosis

* Prominent orbital fat or facial anatomy may simulate proptosis.
* No inflammation or extraocular muscle involvement.

**Epidemiology data**

The prevalence of thyroid eye disease is approximately 50% among GD patients in the Caucasian population. The annual incidence is 16.0 in 100,000 for females and 2.9 in 100,000 for males in the US population.

**Risk Factors**

1. Ethnicity: The African-American population exhibits the maximal risk, followed by the White race and Asian populations.
2. Age: TED shows a bimodal peak incidence. It occurs in age groups of 40 to 44 years and 60 to 64 years in females, and ages of 45 to 49 years and 65 to 69 years in males. It is more severe in older patients with higher chances of restrictive myopathy and dysthyroid optic neuropathy (DON).
3. Gender: There is a female preponderance due to a higher risk of autoimmune diseases. Males cases have more severe ocular involvement and worse outcomes.
4. Genetics: CTLA-4, HLA-DRB-1, and TNF-a genes - are most often associated with TED.
5. Systemic associations: Autoimmune disorders like pernicious anemia, systemic lupus erythematosus, Addison’s disease, vitiligo, coeliac disease, and rheumatoid arthritis - a higher risk of TED.
6. Environmental factors: Smoking is strongly associated with the TED incidence.
7. Dysthyroid status: At the time of diagnosis, 90% of TED cases are hyperthyroid, 6% euthyroid, 3% have Hashimoto thyroiditis, and 1% are hypothyroid.
8. Radioactive iodine therapy (RAIT): causes exacerbation in 24% of TED cases.
9. Stress: Psychological stress can aggravate TED by rebound immune hyperactivity following prolonged corticosteroid-induced immune suppression.
10. Pregnancy: New onset or worsening of TED occurs in 30% of GD cases in the postpartum period.
11. Others: Trauma can be a stimulus for activating an autoimmune cascade in orbit. High serum cholesterol may also be a risk factor for TED.

REFERENCES

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**Trachoma**

**Definition and description**

**Trachoma (truh-KOH-muh)** is a bacterial infection that affects your eyes. It's caused by the bacterium Chlamydia trachomatis. Trachoma is contagious, spreading through contact with the eyes, eyelids, and nose or throat secretions of infected people. It can also be passed on by handling infected items, such as handkerchiefs.

At first, trachoma may cause mild itching and irritation of your eyes and eyelids. Then you may notice swollen eyelids and pus draining from the eyes. Untreated trachoma can lead to blindness.

Trachoma is the leading preventable cause of blindness worldwide. Most trachoma cases occur in poor areas of Africa, where 85% of people with active disease reside. In areas where trachoma is prevalent, infection rates among children under 5 can be 60% or more.

Early treatment may help prevent trachoma complications.

**Causes**

Trachoma is caused by certain subtypes of Chlamydia trachomatis, a bacterium that can also cause the sexually transmitted infection chlamydia.

Trachoma spreads through contact with discharge from the eyes or nose of an infected person. Hands, clothing, towels and insects can all be routes for transmission. In developing countries, eye-seeking flies also are a means of transmission.

## **Risk factors**

Factors that increase your risk of contracting trachoma include:

* Crowded living conditions. People living in close contact are at greater risk of spreading infection.
* Poor sanitation. Poor sanitary conditions, inadequate access to water, and lack of hygiene, such as unclean faces or hands, help spread the disease.
* Age. In areas where the disease is active, it's most common in children ages 4 to 6.
* Sex. In some areas, women's rate of contracting the disease is two to six times higher than that of men. This may be attributed to the fact that women have more contact with children, who are the primary reservoir of infection.
* Flies. People living in areas with problems controlling the fly population may be more susceptible to infection

**Signs and symptoms**

Signs and symptoms of trachoma usually affect both eyes and may include:

* Mild itching and irritation of the eyes and eyelids
* Eye discharge containing mucus or pus
* Eyelid swelling
* Light sensitivity (photophobia)
* Eye pain
* Eye redness
* Vision loss

Young children are particularly susceptible to infection. But the disease progresses slowly, and the more painful symptoms may not emerge until adulthood.

Five stages in the development of trachoma:

* Inflammation — follicular. The early infection has five or more follicles — small bumps that contain lymphocytes, a type of white blood cell — visible with magnification on the inner surface of your upper eyelid (conjunctiva).
* Inflammation — intense. In this stage, your eye is now highly infectious and becomes irritated, with a thickening or swelling of the upper eyelid.
* Eyelid scarring. Repeated infections lead to scarring of the inner eyelid. The scars often appear as white lines when examined with magnification. Your eyelid may become distorted and may turn in (entropion).
* In-turned eyelashes (trichiasis). The scarred inner lining of your eyelid continues to deform, causing your lashes to turn in so that they rub on and scratch the transparent outer surface of your eye (cornea).
* Corneal clouding (opacity). The cornea becomes affected by an inflammation that is most commonly seen under your upper lid. Continuous inflammation compounded by scratching from the in-turned lashes leads to clouding of the cornea.

All the signs of trachoma are more severe in your upper lid than in your lower lid. Without intervention, a disease process that begins in childhood can continue to advance into adulthood.

**Diagnosis**

## Your doctor can diagnose trachoma through a physical examination or by sending a sample of bacteria from your eyes to a laboratory for testing. But lab tests aren't always available in places where trachoma is common.

## **Treatment**

## Trachoma treatment options depend on the stage of the disease.

### Medications

## In the early stages of trachoma, treatment with antibiotics alone may be enough to eliminate the infection. Your doctor may prescribe tetracycline eye ointment or oral azithromycin (Zithromax). Azithromycin appears to be more effective than tetracycline, but it's more expensive.

## The World Health Organization (WHO) recommends giving antibiotics to an entire community when more than 10% of children have been affected by trachoma. The goal of this guideline is to treat anyone who has been exposed to trachoma and reduce the spread of trachoma.

### Surgery

## Treatment of later stages of trachoma — including painful eyelid deformities — may require surgery.

## In eyelid rotation surgery (bilamellar tarsal rotation), your doctor makes an incision in your scarred lid and rotates your eyelashes away from your cornea. The procedure limits the progression of corneal scarring and may help prevent further loss of vision.

## If your cornea has become clouded enough to seriously impair your vision, corneal transplantation may be an option that could improve vision.

## You may have a procedure to remove eyelashes (epilation) in some cases. This procedure may need to be done repeatedly.

## **Prevention**

If you've been treated for trachoma with antibiotics or surgery, reinfection is always a concern. For your protection and for the safety of others, be sure that family members or others you live with are screened and, if necessary, treated for trachoma.

Trachoma can occur worldwide but is more common in Africa, Asia, Latin America, the Middle East and the Pacific Rim. When in regions where trachoma is common, take extra care in practicing good hygiene, which can help prevent infection.

Proper hygiene practices include:

* Face washing and hand-washing. Keeping faces and hands clean may help break the cycle of reinfection.
* Fly control. Reducing fly populations can help eliminate a source of transmission.
* Proper waste management. Properly disposing of animal and human waste can reduce breeding grounds for flies.
* Improved access to water. Having a fresh water source nearby can help improve hygienic conditions.

No trachoma vaccine is available, but prevention is possible. The WHO has developed a strategy to prevent trachoma, with the goal of eliminating it by 2020. While the goal hasn't been entirely achieved, trachoma cases have declined sharply. The strategy, titled SAFE, involves:

* Surgery to treat advanced forms of trachoma
* Antibiotics to treat and prevent the infection
* Facial cleanliness
* Environmental improvements, particularly in water, sanitation and fly control

## **Complications**

One episode of trachoma caused by Chlamydia trachomatis is easily treated with early detection and use of antibiotics. Repeated or secondary infections can lead to complications, including:

* Scarring of the inner eyelid
* **Eyelid deformities, such as an inward-folding eyelid (entropion) or ingrown eyelashes (trichiasis), which can scratch the cornea**
* **Corneal scarring or cloudiness**
* **Partial or complete vision loss**

**When to see a doctor / red flag**

Call your doctor if you or your child has itchy or irritated eyes or discharge from the eyes, especially if you live in or recently traveled to an area where trachoma is common. Trachoma is a contagious condition. Treating it as soon as possible helps prevent serious infection.

## **Differential Diagnoses**

* Allergic Conjunctivitis
* Bacterial Conjunctivitis (Pink Eye)
* Neonatal Conjunctivitis (Ophthalmia Neonatorum)
* Trichiasis
* Viral Conjunctivitis (Pink Eye)

## **EPIDEMIOLOGY**

Trachoma is hyperendemic in many of the poorest and most rural areas of Africa, Central and South America, Asia, Australia and the Middle East.

It is responsible for the blindness or visual impairment of about 1.9 million people. It causes about 1.4% of all blindness worldwide.

Overall, Africa remains the most affected continent and the one with the most intensive control efforts.

As of 21 October 2024, 21 countries – Benin, Cambodia, China, Gambia, Islamic Republic of Iran, Lao People’s Democratic Republic, Ghana, India, Iraq, Malawi, Mali, Mexico, Morocco, Myanmar, Nepal, Oman, Pakistan, Saudi Arabia, Togo, Vanuatu and Viet Nam – had been validated by WHO as having eliminated trachoma as a public health problem.

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**UVEITIS**

**Definition and description**

Uveitis: This is caused by an inflammation of the uvea, the vascular layer (middle layer) inside the eye, which causes redness, pain, and vision problems.

Uveitis is a form of eye inflammation. It affects the middle layer of tissue in the eye wall (uvea).

Uveitis (u-vee-I-tis) warning signs often come on suddenly and get worse quickly. They include eye redness, pain and blurred vision. The condition can affect one or both eyes, and it can affect people of all ages, even children.

Possible causes of uveitis are infection, injury, or an autoimmune or inflammatory disease. Many times a cause can't be identified.

Uveitis can be serious, leading to permanent vision loss. Early diagnosis and treatment are important to prevent complications and preserve your vision.

## 

## **Causes**

In about half of all cases, the specific cause of uveitis isn't clear, and the disorder may be considered an autoimmune disease that only affects the eye or eyes. If a cause can be determined, it may be one of the following:

* An autoimmune or inflammatory disorder that affects other parts of the body, such as sarcoidosis, systemic lupus erythematosus or Crohn's disease.
* Ankylosing spondylitis, a type of inflammatory disease that can cause some of the bones in the spine to fuse, leading to back pain. Uveitis is one of the most common complications of ankylosing spondylitis.
* An infection, such as cat-scratch disease, herpes zoster, syphilis, toxoplasmosis or tuberculosis.
* Medication side effects.
* Eye injury or surgery.
* Very rarely, a cancer that affects the eye, such as lymphoma.

**Risk factors**

People with changes in certain genes may be more likely to develop uveitis. Cigarette smoking has been associated with more difficult to control uveitis

**Symptoms**

The signs, symptoms and characteristics of uveitis may include:

* Eye redness.
* Eye pain.
* Light sensitivity.
* Blurred vision.
* Dark, floating spots in your field of vision (floaters).
* Decreased vision.

Symptoms may occur suddenly and get worse quickly, though in some cases, they develop gradually. They may affect one or both eyes. Occasionally, there are no symptoms, and signs of uveitis are observed on a routine eye exam.

The uvea is the middle layer of tissue in the wall of the eye. It consists of the iris, the ciliary body and the choroid. When you look at your eye in the mirror, you will see the white part of the eye (sclera) and the colored part of the eye (iris).

The iris is located inside the front of the eye. The ciliary body is a structure behind the iris. The choroid is a layer of blood vessels between the retina and the sclera. The retina lines the inside of the back of the eye, like wallpaper. The inside of the back of the eye is filled with a gel-like liquid called vitreous.

The type of uveitis you have depends on which part or parts of the eye are inflamed:

* Anterior uveitis affects the inside of the front of your eye (between the cornea and the iris) and the ciliary body. It is also called iritis and is the most common type of uveitis.
* Intermediate uveitis affects the retina and blood vessels just behind the lens (pars plana) as well as the gel in the center of the eye (vitreous).
* Posterior uveitis affects a layer on the inside of the back of your eye, either the retina or the choroid.
* Panuveitis occurs when all layers of the uvea are inflamed, from the front to the back of your eye.

## **Diagnosis**

When you visit an eye specialist (ophthalmologist), they will likely conduct a complete eye exam and gather a thorough health history. The eye examination usually involves the following:

* **Assessment of vision** (with your glasses if you normally wear them) and the response of your pupils to light.
* **Tonometry.** A tonometry exam measures the pressure inside your eye (intraocular pressure). Numbing eyedrops may be used for this test.
* **A slit-lamp examination.** A slit lamp is a microscope that magnifies and illuminates the front of your eye with an intense line of light. This evaluation is necessary to identify microscopic inflammatory cells in the front of the eye.
* **Ophthalmoscopy.** Also known as funduscopy, this exam involves widening (dilating) the pupil with eye drops and shining a bright light into the eye to examine the back of the eye.

Your doctor also may recommend:

* **Color photography** of the inside of the eye (retina).
* **Optical coherence tomography (OCT) imaging.** This test maps the retina and choroid to reveal swelling in these layers.
* **Fluorescein angiography or indocyanine green angiography.** These tests require placement of an intravenous (IV) catheter in a vein in your arm in order to give a dye. This dye will reach the blood vessels in the eyes and allow photographs of swollen blood vessels inside the eyes.
* **Analysis of aqueous or vitreous fluid** from the eye.
* **Blood tests.**
* **Imaging tests,** radiography, Computerized tomography (CT) or Magnetic resonance imaging (MRI) scans.

If the ophthalmologist thinks an underlying condition may be the cause of your uveitis, you may be referred to another doctor for a general medical examination and laboratory tests.

Sometimes, it's difficult to find a specific cause for uveitis. Even if a specific cause is not identified, uveitis can still be treated successfully. In the majority of cases, identifying a cause for the uveitis does not lead to a cure. It is still necessary to use some form of treatment to control the swelling.

**Treatment**

If uveitis is caused by an underlying condition, treatment may focus on that specific condition. Usually the treatment for uveitis is the same regardless of the cause, as long as the cause is not infectious. The goal of treatment is to reduce the swelling in your eye, as well as in other parts of the body, if present. In some cases, treatment may be necessary for months to years. Several treatment options are available.

### **Medications**

* **Drugs that reduce inflammation.** Your doctor may first prescribe eye drops with an anti-inflammatory medication, such as a corticosteroid. Eye drops are usually not enough to treat inflammation beyond the front of the eye, so a corticosteroid injection in or around the eye or corticosteroid tablets (taken by mouth) may be necessary.
* **Drugs that control spasms.** Eye drops that widen (dilate) the pupil may be prescribed to control spasms in the iris and ciliary body, which can help relieve eye pain.
* **Drugs that fight bacteria or viruses.** If uveitis is caused by an infection, your doctor may prescribe antibiotics, antiviral medications or other medicines, with or without corticosteroids, to bring the infection under control.
* **Drugs that affect the immune system or destroy cells.** You may need immunosuppressive drugs if your uveitis affects both eyes, doesn't respond well to corticosteroids or becomes severe enough to threaten your vision.

Some of these medicines can have serious eye-related side effects, such as glaucoma and cataracts. Medicine by mouth or injection can have side effects in other parts of the body outside the eyes. You may need to visit your doctor frequently for follow-up examinations and blood tests.

### **Surgical or other procedures**

* **Vitrectomy.** Surgery to remove some of the vitreous in your eye is rarely used to diagnose or manage the condition.
* **A medication-releasing implant.** For people with difficult-to-treat posterior uveitis, a device that's implanted in the eye may be an option. This device slowly releases corticosteroid into the eye for months or years depending on the implant used.  
  If people have not had cataract surgery, this treatment usually causes cataracts to develop. Also, up to 30% of patients will require treatment or monitoring for elevated eye pressure or glaucoma.

The speed of your recovery depends in part on the type of uveitis you have and how bad your symptoms are. Uveitis that affects the back of your eye (posterior uveitis or panuveitis, including retinitis or choroiditis) tends to heal more slowly than uveitis in the front of the eye (anterior uveitis or iritis). Severe inflammation takes longer to clear up than mild inflammation does.

Uveitis can come back. Make an appointment with your doctor if any of your symptoms reappear or worsen.

**PROGNOSIS**

In most cases, the prognosis of uveitis is good assuming early detection and proper treatment. While a majority of patients will develop an ocular complication, appropriate treatment and surgery, if needed, make permanent vision loss much less likely.. Identifying the underlying cause of uveitis is also important due to significant morbidity and mortality associated with some specific systemic diseases that can cause uveitis.

**Possible complications**

Left untreated, uveitis can cause complications, including:

* Retinal swelling (macular edema).
* Retinal scarring.
* Glaucoma.
* Cataracts.
* Optic nerve damage.
* Retinal detachment.
* Permanent vision loss.

**When to see a doctor / red flag**

Contact your doctor if you think you have the warning signs of uveitis. He or she may refer you to an eye specialist (ophthalmologist). If you're having significant eye pain and unexpected vision problems, seek immediate medical attention.

**DIFFERENTIAL DIAGNOSIS**

There are several other causes for a painful, red eye with vision changes/loss that are frequently encountered in the primary care and ER setting. It is important to keep a broad differential when evaluating an eye complaint as there can be significant overlap with a patient's symptoms. The following causes of a red, painful eye require special skills that must be used in each case to differentiate them. These special skills include visual acuity testing, IOP measurements, slit lamp biomicroscopy, and fluorescein staining.

Conjunctivitis can present as a painful red eye with photophobia. Bacterial conjunctivitis can present with purulent discharge which is not present in uveitis. Patients with viral or allergic conjunctivitis will usually have serous discharge; however, they can also be without discharge. Follicles of the palpebral conjunctiva and the lack of anterior chamber inflammation help make the diagnosis of conjunctivitis. Chemosis (swelling of the conjunctiva, is also a finding found in conjunctivitis but not in uveitis.

Keratitis is significant inflammation of the cornea. It frequently presents with pain, photophobia, and perilimbic injection and is most commonly seen in patients with a history of contact lens use. In keratitis, the cornea is often clouded if not frankly opaque (i.e. corneal ulcer, with ring infiltrate), which is not seen in uveitis. Also, the pupil is usually normal in shape in keratitis but may be constricted in anterior uveitis due to posterior synechiea formation . Fluoroscein dye will identify a brightly staining epithelial defect when viewed under a cobalt light, same patient as in 1f with epithelial defect identified by fluorescein dye). The presence of stromal whitening (corneal infiltrate, that is seen with keratitis is not seen with corneal abrasions.

Acute angle closure glaucoma can present as a painful red eye with a change in vision. The patient may complain of a unilateral headache and maybe nauseous even to the point of vomiting. The affected pupil is often mid-dilated and poorly reactive to light. The IOP will be significantly increased in acute angle closure glaucoma and is typically seen in most causes of uveitis. However, an elevated IOP in the setting of a unilateral anterior uveitis is seen with herpes infections, so a slit lamp examination specifically looking for anterior chamber cell and keratic precipitates (punctate spots on corneal endothelium) should be performed in each case to properly evaluate and treat the patient. The cornea may also be cloudy due to the acute rise in IOP in angle closure glaucoma.

A corneal abrasion will often cause severe eye pain, photophobia, and tearing. Patients will usually endorse a history of trauma to the affected eye. A drop of proparacaine at time of evaluation will nearly, if not completely, treat the pain. This sensitivity to topical anesthesia is not a feature of uveitis of any type. Examination with fluorescein will identify the abrasion as a brightly staining lesion of the cornea and unless infected, will not likely have a stromal infiltrate/whitening.

A panuveitis such as seen in acute retinal necrosis from herpes infections and endophthalmitis causes severe inflammation resulting in a red, painful eye. A hypopyn may be found in the anterior chamber. As discussed elsewhere, this finding should raise alarm. A dilated eye exam is required to identify retinal whitening seen in acute retinal necrosis and significant vitritis in endophthalmitis, and as such, likely require an ophthalmology consultation. If these diagnoses are suspected, an emergent ophthalmology consult should be placed as both are ophthalmic emergencies with earlier treatment resulting in better outcomes.

**Most common causes of a red, painful eye:**

* Anterior uveitis
* Keratitis
* Corneal abrasion
* Acute angle closure glaucoma
* Conjunctivitis
* Scleritis

**EPIDEMIOLOGY**

Uveitis can affect people of all ages and can vary significantly by geographic location and age of the patient . In a study done from 2006 to 2007, the incidence of uveitis was 24.9 cases per 100,000 persons. Prevalence rates for 2006 and 2007 came in at 57.5 and 58 respectively per 100,000 persons . There was no difference in the incidence rate between men and women, but women had a higher prevalence . Anterior uveitis is the most prevalent form, accounting for approximately 50% of uveitis cases, while posterior uveitis is the least common. Ongoing inflammation seen in untreated uveitis and complications related to this uncontrolled inflammation are estimated to be responsible for approximately 10% of the blindness in the United States.

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**VITREOMACULAR TRACTION SYNDROME**

**Definition and description**

The vitreous humor is a transparent, gel-like material that fills the space within the eye between the lens and the retina. The vitreous is encapsulated in a thin shell called the vitreous cortex, and the cortex in young, healthy eyes is usually sealed to the retina.

As the eye ages, or in certain pathologic conditions, the vitreous cortex can pull away from the retina, leading to a condition known as posterior vitreous detachment (PVD). This detachment usually occurs as part of the normal aging process.

There are instances where a PVD is incomplete, leaving the vitreous partially attached to the retina, and causing tractional (pulling) forces that can cause anatomical damage. The resulting condition is called vitreomacular traction (VMT) syndrome.

VMT syndrome can lead to different maculopathies or disorders in the macular area (at the center of the retina), such as full- or partial-thickness **macular holes**, **epiretinal membranes**, and **cystoid macular edema**. These disorders are often associated with reduced sharpness of vision (visual acuity) or other visual complications.

**The most common symptoms experienced by patients with VMT syndrome are:**

* Decreased sharpness of vision
* Photopsia, when a person sees flashes of light in the eye
* Micropsia, when objects appear smaller than their actual size
* Metamorphopsia, when vision is distorted to make a grid of straight lines appear wavy or blank

Some of these symptoms can be mild and develop slowly; however, chronic tractional effects can lead to continued visual loss if left untreated. In some cases, a distortion of a visual picture could be experienced without necessarily having a reduction in sharpness of vision.

**Causes**

Age-related degeneration of the gel-like vitreous humor leads to the formation of pockets of fluid within the vitreous, causing contraction and loss of volume. The separation of the vitreous gel from the retina occurs as a result of the gel becoming liquid (liquefaction) and the continuous anterior-posterior (front-back) and tractional forces stretching on the **macula** over time.

Weakening of the attachments of the vitreous cortex and the **internal limiting membrane (ILM)** of the retina could also lead to partial detachment of the posterior hyaloid membrane, leading to PVD and potentially VMT.

## **Risk factors**

VMT syndrome is most common in older adults and women due to age-related vitreous changes and vitreous liquefaction associated with declining postmenopausal estrogen levels, respectively.

* Other risk factors include:
* High myopia (extreme nearsightedness)
* Exudative (wet) age-related macular degeneration
* Diabetic macular edema
* Retinal vein occlusion
* Diabetic retinopathy

**Diagnostic testing**

**Optical coherence tomography (OCT)** is a commonly used and recommended method to noninvasively identify and monitor VMT syndrome. This technology captures cross-sectional images of the retinal layers, including the surface, and allows physicians to evaluate the degree to which vitreomacular tractional forces are distorting the retinal structure.

A **dynamic B-scan ultrasound** examination could also be performed to provide a detailed evaluation of the vitreoretinal interface.

## **Treatment and prognosis**

There are currently 4 main options for treating VMT syndrome.

1. Watchful waiting and regular monitoring with OCT is often used for patients whose symptoms do not warrant active intervention. Some cases of VMT may spontaneously resolve.
2. For patients whose symptoms are severe enough to require intervention, **pars plana vitrectomy** **surgery** is one treatment option. The procedure involves the manual release of vitreous attachment and alleviation of traction, but it is invasive and inconvenient to most patients. Therefore vitrectomy is reserved for patients who are at risk for severe visual disturbances and/or central blindness. Some studies have shown that shorter duration of symptoms results in better prognosis when the surgical treatment is employed.
3. Ocriplasmin (Jetrea®), a recombinant truncated form of human plasmin, is a pharmacologic option for treating VMT syndrome. Clinical trials demonstrated the efficacy and safety of a single intravitreal injection of ocriplasmin for the treatment of patients with symptomatic vitreomacular adhesion, and/or vitreomacular traction. Ocriplasmin is therefore a treatment option for some patients who have vitreomacular traction but who are not candidates for surgery.
4. Pneumatic vitreolysis - a small gas bubble is injected into the eye in the office. Afterward, the patient will look down several times per hour for a day or two to try to get the gas bubble to sever the adhesion between the vitreous and the macula.

* Most patients with VMT maintain good visual acuity in the affected eye, even if treatment is required

.Epidemiology of Vitreomacular Traction (VMT)

* Prevalence and Incidence:  
  The prevalence of isolated VMT syndrome is approximately 22.5 per 100,000 population, with an annual incidence of about 0.6 per 100,000 population. However, when considering symptomatic vitreomacular adhesion (sVMA), which includes VMT and early-stage macular holes (MH), the prevalence is higher at about 1,365 cases per 100,000 population, with an incidence rate of 6.96 new cases per 100,000 per year.
* Population Impact:  
  For example, in Germany, with a population of roughly 82 million, this translates to approximately 1,119,300 prevalent cases of sVMA and about 5,700 new cases annually.
* Age and Detection:  
  VMT and related vitreoretinal interface disorders are more common in older individuals and are increasingly diagnosed due to the widespread use of optical coherence tomography (OCT). OCT allows earlier and more sensitive detection, including asymptomatic cases that may not progress to clinical intervention.
* Associated Conditions:  
  The prevalence and incidence of VMT are higher when associated with other macular diseases such as diabetic retinopathy, diabetic macular edema, and age-related macular degeneration.
* Clinical vs Population Data:  
  There is a discrepancy between population-based prevalence and clinical incidence rates. Many cases detected in population studies may be asymptomatic and do not present to clinics, which likely explains lower clinical incidence compared to prevalence estimates

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<https://www.asrs.org/patients/retinal-diseases/12/vitreomacular-traction-syndrome>

**WOLFRAM SYNDROME**

*ALTERNATIVE NAMES:* Wolfram syndrome is also known by several alternative names. One of the most common is “DIDMOAD”, which stands for “diabetes insipidus”, “diabetes mellitus”, “optic atrophy”, and “deafness”.

Another name for the syndrome is the “diabetes insipidus-diabetes mellitus-optic atrophy-deafness syndrome”.

**DEFINITION / DESCRIPTION**

Wolfram syndrome is a rare genetic disease. It’s a progressive, neurodegenerative disorder that damages your brain and other tissues in your body. A series of symptoms usually appear during childhood and into adulthood. Diabetes and vision changes before age 15 are usually the first symptoms. Eventually, impaired brain function can lead to early death.

**Types of Wolfram syndrome**

Healthcare providers have identified two genes involved in Wolfram syndrome. Genes are sequences of DNA that carry genetic information.

People with Wolfram syndrome have changes (mutations) in their genes. Healthcare providers classify Wolfram syndrome based on the affected genes:

* Wolfram syndrome type 1 is the result of a mutation of the *WFS1* gene.
* Wolfram syndrome type 2 is the result of a mutation of the *WFS2* (*CISD2*) gene.

**How is Wolfram syndrome inherited?**

To pass on Wolfram syndrome, usually both biological parents must carry the same gene mutation. But in some cases, a person can inherit Wolfram syndrome type 1 when only one parent has the mutation.

Because Wolfram syndrome is so rare, healthcare providers don’t know exactly how often it occurs. One study estimated that Wolfram syndrome affects 1 in 770,000 people in the United Kingdom. Studies from other countries suggest it may be more common in areas where people who are close relatives marry and have children.

Wolfram syndrome type 2 is extremely rare. Healthcare providers have reported cases in only a few families worldwide.

**SIGNS / SYMPTOMS**

***What are the four most common features of Wolfram syndrome type 1?***

Symptoms of Wolfram syndrome type 1 may vary from person to person. But often, they appear in a predictable order during childhood and adolescence. This is the typical sequence and average age that symptoms appear:

1. ***Diabetes mellitus (age 6):*** Diabetes mellitus is a problem with your body’s ability to absorb sugar (glucose) from the food you eat. Normally, your pancreas makes insulin, which helps your cells absorb sugars (glucose) from your bloodstream. If you don’t make enough insulin or if your cells don’t respond to insulin, your blood sugar can rise too high. Wolfram syndrome-related diabetes is similar to Type 1 diabetes, but it’s not an autoimmune disease. Diabetes symptoms include frequent urination, increased thirst, blurred vision and unexplained weight loss.
2. ***Optic atrophy (age 11):*** Optic atrophy is the degeneration of your optic nerve, which carries signals from your eyes to your brain. Symptoms include blurred, dulled or reduced peripheral (side) vision.
3. ***Sensorineural hearing loss (age 13):*** Sensorineural hearing loss occurs due to damage in your inner ear. This type of hearing loss usually gets worse as you get older and can lead to deafness.
4. ***Diabetes insipidus (age 14):*** Diabetes insipidus isn’t related to diabetes mellitus. It’s an issue with the production of an antidiuretic hormone that controls the amount of water in your urine (pee). People with diabetes insipidus have large amounts of watery urine. This excess urination can cause dehydration, electrolyte disturbance, weakness, dry mouth and constipation.

An outdated name for Wolfram syndrome is DIDMOAD. This acronym incorporates the primary symptoms:

* Diabetes insipidus (DI).
* Diabetes mellitus (DM).
* Optic atrophy (OA).
* Deafness (D).

**Symptoms of Wolfram syndrome type 1**

Less common symptoms include:

* Hormone disorders: Hypopituitarism, low sex drive (hypogonadism), growth delays and delayed start of menstrual periods.
* Neurological symptoms: Problems with movement and coordination (ataxia), dementia, headaches, central sleep apnea, epilepsy and decreased sense of taste and smell.
* Psychiatric conditions: Depression, anxiety, panic attacks, mood swings and aggressive behavior.
* Urinary tract abnormalities: Urinary tract infections, urinary incontinence and incomplete emptying of your bladder.

**Symptoms of Wolfram syndrome type 2**

Symptoms of Wolfram syndrome type 2 are similar to type 1, but may also include:

* Abnormal bleeding.
* Gastrointestinal ulcers.

People with Wolfram syndrome type 2 usually don’t have diabetes insipidus or psychiatric conditions.

**CAUSES**

Wolfram syndrome is primarily caused by mutations in the WFS1 gene, which encodes a protein called wolframin. These mutations are responsible for more than 90% of Wolfram syndrome type 1 cases.

The WFS1 gene is located on chromosome 4p and plays a role in regulating calcium balance within cells, which is crucial for various cellular functions. Mutations in the WFS1 gene can lead to a variety of symptoms, including diabetes mellitus, optic atrophy, diabetes insipidus, and sensorineural hearing loss.

Additionally, mutations in the CISD2 gene can cause Wolfram syndrome 2 (WS2), which is characterized by the absence of diabetes insipidus and psychiatric disorders, along with other features such as bleeding upper intestinal ulcers and defective platelet aggregation.

Variants (also known as mutations) in the *WFS1* gene cause more than 90 percent of Wolfram syndrome type 1 cases. This gene provides instructions for producing a protein called wolframin that is thought to regulate the amount of calcium in cells. A proper calcium balance is important for many different cellular functions, including cell-to-cell communication, the tensing (contraction) of muscles, and protein processing. The wolframin protein is found in many different tissues, such as the pancreas, brain, heart, bones, muscles, lung, liver, and kidneys. Within cells, wolframin is located in the membrane of a cell structure called the endoplasmic reticulum that is involved in protein production, processing, and transport. Wolframin's function is important in the pancreas, where the protein is thought to help process a protein called proinsulin into the mature hormone insulin. This hormone helps control blood glucose levels.

*WFS1* gene variants lead to the production of a wolframin protein that has reduced or absent function. As a result, calcium levels within cells are not regulated and the endoplasmic reticulum does not work correctly. When the endoplasmic reticulum does not have enough functional wolframin, the cell triggers its own cell death (apoptosis). The death of cells in the pancreas, specifically cells that make insulin (beta cells), causes diabetes mellitus in people with Wolfram syndrome. The gradual loss of cells along the optic nerve eventually leads to blindness in affected individuals. The death of cells in other body systems likely causes the various signs and symptoms of Wolfram syndrome type 1.

A certain variant in the *CISD2* gene was found to cause Wolfram syndrome type 2. The *CISD2* gene provides instructions for making a protein that is located in the outer membrane of cell structures called mitochondria. Mitochondria are the energy-producing centers of cells. The exact function of the CISD2 protein is unknown, but it is thought to help keep mitochondria functioning normally.

The *CISD2* gene variant that causes Wolfram syndrome type 2 results in an abnormally small, nonfunctional CISD2 protein. As a result, mitochondria are not properly maintained, and they eventually break down. Since the mitochondria provide energy to cells, the loss of mitochondria results in decreased energy for cells. Cells that do not have enough energy to function will eventually die. Cells with high energy demands such as nerve cells in the brain, eye, or gastrointestinal tract are most susceptible to cell death due to reduced energy. It is unknown why people with *CISD2* gene variants have ulcers and bleeding problems in addition to the usual Wolfram syndrome features.

Some people with Wolfram syndrome do not have an identified variant in either the *WFS1* or *CISD2* gene. The cause of the condition in these individuals is unknown.

**What causes Wolfram syndrome?**

Wolfram syndrome occurs when biological parents pass on changes (mutations) in the *WFS1* or *WFS2* (*CISD2*) genes to their children.

**RISK FACTORS**

Wolfram syndrome is a rare genetic disorder primarily caused by mutations in the WFS1 or WFS2 genes, which are inherited in an autosomal recessive pattern.

This means that both copies of the gene in each cell are altered, and the parents of an individual with Wolfram syndrome each carry one copy of the altered gene but typically do not show symptoms.

Some studies have shown that individuals who have one copy of a WFS1 gene variant may be at increased risk of developing individual features of Wolfram syndrome or related features, such as type 2 diabetes, hearing loss, or psychiatric illness. However, other studies have found no increased risk in such individuals.

Wolfram syndrome caused by variants in the CISD2 gene is also inherited in an autosomal recessive pattern.

In addition, dominant forms of Wolfram syndrome have been described, where only a single copy of a disease-causing gene variant is necessary to cause the disease.

The gene variant can be inherited from either parent or can be the result of a new (de novo) changed gene in the affected individual that is not inherited. The risk for two carrier parents to both pass the gene variant and have an affected child is 25% with each pregnancy.

The risk of having a child who is a carrier like the parents is 50% with each pregnancy. The chance for a child to receive normal genes from both parents is 25%.

It is important to note that while the primary cause of Wolfram syndrome is genetic, the exact mechanisms and additional risk factors are still being studied.

**DIAGNOSIS METHODS AND TESTS**

Diagnosing Wolfram syndrome can be challenging. Healthcare providers may diagnose individual conditions but not make the connection between them. Often, diagnosis occurs after multiple symptoms develop.

If your provider suspects Wolfram syndrome, they’ll recommend genetic testing. This test detects mutations in the *WFS1* and *WFS2* (*CISD2*) genes and can confirm your diagnosis.

The diagnosis of Wolfram syndrome involves a combination of clinical evaluation and genetic testing. Genetic testing is a crucial component in confirming the diagnosis, as it allows for the identification of specific mutations associated with the condition. The primary genetic locus associated with Wolfram syndrome is the WFS1 gene, and Sanger sequencing of this gene typically confirms the diagnosis.

Most patients exhibit recessive mutations in WFS1, meaning they inherit two copies of the mutated gene, one from each parent. However, some dominant mutations, such as H313Y, have been identified, where one copy of the mutated gene can cause the disorder.

In addition to WFS1, mutations in the CISD2 (CDGSH iron-sulfur domain-containing protein 2) gene have been linked to autosomal recessive Wolfram syndrome 2 (WS2).

For patients who do not carry mutations in the WFS1 gene, Sanger sequencing-based genetic testing of the WFS2 gene is performed.

Exome sequencing and genome sequencing-based diagnostic methods are also being developed for Wolfram syndrome and Wolfram-related disorders.

Clinical evaluation is also essential, as Wolfram syndrome should be considered in individuals with diabetes mellitus and optic atrophy, those with low-frequency sensorineural hearing loss, or those with diabetes mellitus or optic atrophy in addition to hearing loss, diabetes insipidus, bladder dysfunction, or loss of sense of smell.

Early diagnosis is imperative to enable proper prognostication, prevent complications, and start available treatments.

**MANAGEMENT AND TREATMENT OPTIONS**

Currently, there aren’t any standard treatments to stop or slow the progression of Wolfram syndrome. Treatment focuses on managing related symptoms. For example, healthcare providers treat diabetes mellitus with insulin to help manage blood sugar levels. Hearing aids can help people with hearing loss.

**Are other treatments under investigation?**

Researchers are studying therapies that may improve the outlook for people with Wolfram syndrome. Leading treatment strategies include:

* Drugs to reduce cell damage caused by disruption of proteins.
* Gene therapy to repair or replace mutated *WFS1* and *WFS2* (*CISD2*) genes.
* Regenerative therapy to heal or replace damaged tissues.

Some treatments are now available through clinical trials. Others are still in development. Talk to your healthcare provider about emerging treatments and whether they might be an option for you or your child.

**OUTLOOK / PROGNOSIS**

People with Wolfram syndrome have a poor prognosis. In one study of 45 patients, life expectancy ranged from 25 to 49 years with an average of 30. The most common cause of death was due to deterioration of the brainstem that controls vital functions like breathing and heart rate.

Improved diagnosis and management and the prospect of new therapies are improving the outlook.

**PREVENTION TIPS**

You can’t prevent genetic conditions such as Wolfram syndrome.

Wolfram syndrome is a rare genetic disorder with no known preventive measures due to its hereditary nature. However, early diagnosis and management can help in reducing complications and improving the quality of life for affected individuals.

Regular medical check-ups, especially for those with a family history of the syndrome, are crucial for early detection.

Additionally, maintaining a healthy lifestyle, including a balanced diet and regular exercise, can help manage some of the symptoms associated with Wolfram syndrome, such as diabetes mellitus.

It is also important to monitor and manage the symptoms promptly to prevent further complications. Genetic counseling is recommended for families with a history of Wolfram syndrome to understand the risks and options for future pregnancies.

**POSSIBLE COMPLICATIONS**

Wolfram syndrome is a rare autosomal-recessive genetic disorder that can lead to a variety of complications, primarily affecting the endocrine, neurological, and sensory systems. Some of the possible complications include:

* Diabetes mellitus: This is a key feature of Wolfram syndrome, often appearing in childhood. It results from the improper control of glucose due to the lack of insulin.
* Optic atrophy: This leads to a gradual loss of vision as the nerve that connects the eye to the brain wastes away.
* Deafness: Sensorineural hearing loss is common, which can be partial or complete and may occur at any age.
* Diabetes insipidus: This condition affects the kidneys' ability to conserve water, leading to excessive urination and thirst.
* Neurological complications: These may include problems with movement and coordination (ataxia), dementia, headaches, central sleep apnea, epilepsy, and decreased sense of taste and smell.
* Psychiatric conditions: Depression, anxiety, panic attacks, mood swings, and aggressive behavior can occur.
* Urinary tract abnormalities: These may include urinary tract infections, urinary incontinence, and incomplete emptying of the bladder.
* Neurodegeneration: This can lead to brain stem atrophy, which is a major cause of death in Wolfram syndrome patients, typically due to respiratory failure.
* Hearing loss: This is due to a loss of sound perception transmitted by nerves from the ear to the brain.
* Genitourinary problems: These can include issues with the bladder not emptying properly, leading to frequent urination.
* Mitochondrial dysfunction: This can lead to premature aging, mitophagy, and other cellular dysfunctions.
* Increased risk of bleeding and peptic ulcers: These are more commonly associated with Wolfram syndrome type 2.

These complications highlight the need for a multidisciplinary approach to manage the various aspects of Wolfram syndrome.

**WHEN TO SEE A DOCTOR / RED FLAG**

When should I talk to a healthcare provider about genetic testing?

If you have a family history of Wolfram syndrome, talk to your provider about genetic testing. With a simple blood or saliva test, you can:

* Determine your risk of having a child with the disorder.
* Learn whether your child has Wolfram syndrome before they develop symptoms.

**DIFFERENTIAL DIAGNOSIS**

*Differential Diagnosis of Wolfram Syndrome (WFS)*

Wolfram syndrome, a rare genetic disorder characterized primarily by juvenile-onset diabetes mellitus and optic atrophy, has a broad differential diagnosis due to overlapping features with other genetic, mitochondrial, and neurodegenerative disorders. Key differential diagnoses include:

* Mitochondrial Disorders
  + Maternally Inherited Diabetes and Deafness (MIDD)
  + Leber Hereditary Optic Neuropathy (LHON)
  + Kearns-Sayre Syndrome
  + Mohr-Tranebjaerg Syndrome  
    These disorders share features such as diabetes, optic atrophy, and sensorineural hearing loss.
* Autosomal Dominant Optic Atrophy  
  Presents with optic nerve atrophy and may have hearing loss, resembling Wolfram syndrome but with different inheritance patterns.
* Thiamine-Responsive Megaloblastic Anemia (TRMA) Syndrome  
  Includes diabetes and optic atrophy among its features.
* Wolfram-like Syndrome  
  An autosomal dominant disorder with adult-onset diabetes and juvenile optic atrophy, sometimes with hearing impairment.
* Friedreich Ataxia  
  A neurodegenerative disorder that can mimic neurological signs seen in Wolfram syndrome.
* Bardet-Biedl Syndrome and Alström Syndrome  
  Multisystem disorders with overlapping features such as diabetes and sensory deficits.
* X-linked Charcot-Marie-Tooth Disease Type 5 (CMTX5)  
  Characterized by neuropathy, optic atrophy, and hearing loss.
* Deafness-Dystonia-Optic Neuropathy (DDON) Syndrome  
  Shares sensorineural deafness and optic neuropathy features.
* Diabetic Papillopathy  
  Can present with optic nerve head swelling in diabetic patients, potentially confused with optic atrophy.

**RECENT GUIDELINES OR UPDATES**

Wolfram syndrome is a rare genetic disorder characterized by childhood-onset diabetes mellitus, optic atrophy, deafness, diabetes insipidus, and neurological signs.

There are no established treatment guidelines for treating or slowing down the progression of Wolfram Syndrome, and treatment is primarily focused on managing the symptoms.

However, research is ongoing to find more effective treatment strategies, including studies on drugs that can reduce cell damage, gene therapies, and regenerative therapies.

Recent studies have suggested that doses of dantrolene sodium less than 200 mg/day may be safely used in patients without coexisting liver dysfunction or co-ingestion of hepatotoxic medications. There is also a phase 1 clinical trial currently investigating the safety of dantrolene long-term use in WS patients.

Additionally, therapeutic strategies in WS are based on drug repurposing, aiming to stop the progression of the disease by reducing the endoplasmic reticulum stress.

Despite these efforts, there is currently no treatment to delay, halt, or reverse the progression of Wolfram syndrome, highlighting the urgency for innovative therapeutics for this disease.

The prognosis for Wolfram syndrome is very poor, with a median mortality rate of 65% before the age of 35.

**EPIDEMIOLOGY**

The estimated prevalence of Wolfram syndrome (WS) worldwide ranges between about 1 in 770,000 and 1 in 100,000, depending on the population studied. More specifically:

* In the United Kingdom, prevalence is estimated at approximately 1 in 770,000 people.
* In North America, it is higher, around 1 in 100,000.
* In Japan, the prevalence is about 1 in 710,000.
* In Lebanon and certain Sicilian populations, prevalence is notably higher, reaching as high as 1 in 68,000 and even 1 in 54,478 in a small Sicilian district, likely due to higher rates of consanguinity.
* In India, estimates suggest a prevalence near 1 in 805,000.

Wolfram syndrome type 1 (WS1) is the most common form, with juvenile-onset diabetes mellitus and optic atrophy as hallmark features. The carrier frequency of WS1 mutations can be as high as 1% in some populations, with a heterozygous carrier rate of about 1 in 354 in the UK. The syndrome is autosomal recessive.

Among patients with juvenile-onset insulin-dependent diabetes mellitus, the prevalence of Wolfram syndrome varies between approximately 1 in 148 and 1 in 175, indicating it is an important but rare cause of diabetes in youth.

WS2, caused by mutations in a different gene, is much rarer, with only a few families described.

The syndrome is associated with significant morbidity and early mortality, with a median age of death around 39 years, often due to neurological complications.

*REFERENCES*

[Wolfram Syndrome: What It Is, Symptoms & Treatment](https://my.clevelandclinic.org/health/diseases/24044-wolfram-syndrome#overview)

<https://medlineplus.gov/genetics/condition/wolfram-syndrome/#resources>

**UVEAL MELANOMA**

*ALTERNATIVE NAMES:* Uveal melanoma is also known as intraocular melanoma.

**DEFINITION / DESCRIPTION**

Uveal melanoma, sometimes called ocular melanoma, is a type of cancer that affects the uvea, the middle layer of the eye that contains blood vessels, pigment cells (melanocytes), and provides nutrients and oxygen to the retina. It is a rare form of melanoma derived from the melanocytes in the uvea, accounting for only about 5% of all cases of melanoma in the United States.

Uveal melanoma can develop in different parts of the uvea, including the iris, ciliary body, and choroid. Choroidal melanoma is the most common site (90% of all uveal melanomas) followed by ciliary body (6%) and iris (4%) melanomas.

The rate of new cases of uveal melanomas have remained fairly consistent over the last three decades across the world.

**SIGNS / SYMPTOMS**

***Signs and symptoms of uveal melanoma***

Uveal melanoma may not cause any symptoms in its early stages, and it may be detected during a routine eye exam with pupil dilation, which is the best way to screen for uveal melanoma. However, as the tumor grows, it can cause a variety of symptoms, including:

* Blurred vision or loss of vision in one eye
* Dark spots on the iris (the colored part of the eye) - A change in the shape or size of the pupil
* A bulging of the eye
* Flashes of light or floating spots in the vision
* Pain or redness in the eye

It's important to note that these symptoms can also be caused by other eye conditions, so it's important to see an eye doctor if you experience any changes in your vision or eye health. If uveal melanoma is suspected, your doctor may perform additional tests, such as an ultrasound or biopsy, to confirm the diagnosis. Early detection and treatment of uveal melanoma can improve the chances of a successful outcome.

***How common is uveal melanoma?***

Uveal melanoma is a rare form of melanoma, accounting for only about 5% of all cases. About -5000 cases of uveal melanoma are diagnosed each year in the United States.

***How serious is uveal melanoma?***

Uveal melanoma is a serious condition that requires prompt medical attention. While it is a rare form of melanoma, if undiagnosed, it can be aggressive and can spread to other parts of the body, such as the liver (most common organ for uveal melanoma to spread to), lungs, brain, kidney, and bones. In fact, up to 50% of patients with uveal melanoma will develop metastatic disease, which can be life-threatening.

The prognosis for uveal melanoma depends on a number of factors, including the size and location of the tumor, the extent of its spread, and the patient's overall health. In general, smaller tumors that have not spread beyond the eye have a better prognosis than larger tumors or those that have spread to other parts of the body.

Who gets uveal melanoma?

Uveal melanoma is a rare type of cancer that can affect anyone, but it is more common in certain groups of people. Some of the factors that may increase the risk of developing uveal melanoma include:

* Gender: Men have a slightly higher chance of getting uveal melanoma than women.
* Age: Uveal melanoma is most commonly diagnosed in people over the age of 50 and is rarely found in children.
* Race: The risk of uveal melanoma is lowest in Blacks followed by increased rates in Asians, then Hispanics, then non-Hispanics, respectively.
* Physical features: Uveal melanoma is more common in people with light-colored eyes, such as blue or green eyes, and fair skin.
* Genetics: Some inherited genetic mutations have been linked to an increased risk of uveal melanoma which can be diagnosed at a younger age, including mutations in the BAP1 gene.
* Exposure to ultraviolet (UV) radiation: While UV radiation from the sun is a known risk factor for skin cancer, it is not clear if it plays a role in the development of uveal melanoma.

It's important to note that many people who develop uveal melanoma do not have any known risk factors, and the exact cause of the disease is not fully understood. If you are concerned about your risk of developing uveal melanoma, talk to your healthcare provider. They can help you understand your risk factors and recommend appropriate screening tests or preventive measures.

Causes uveal melanoma

The exact cause of uveal melanoma is not fully understood, but it is believed to be related to mutations that occur in the pigment cells of the eye. These mutations can cause the cells to grow and divide uncontrollably, leading to the development of a uveal tumor. The mutations found in uveal melanoma are distinct from those found in other melanoma subtypes.

How is uveal melanoma treated?

The treatment of uveal melanoma depends on several factors, including the size and location of the tumor, as well as the patient's overall health. Treatment options may include:

* Radiation therapy: This is the most common treatment for uveal melanoma. Radiation therapy uses different types of high-energy beams of radiation to destroy cancer cells. It can be delivered externally (external beam radiation therapy) or internally (brachytherapy).
* Surgery: Surgery may be an option for small tumors that have not spread beyond the eye. The goal of surgery is to remove the tumor and a small amount of surrounding tissue.
* Laser therapy: Laser therapy uses a high-energy beam of light to destroy cancer cells. It may be used to treat small tumors that are located in the front of the eye.
* Chemotherapy: Chemotherapy is not commonly used to treat uveal melanoma, but it may be an option for advanced cases that have spread to other parts of the body.
* Immunotherapy: Immunotherapy is a type of treatment that uses the body's immune system to fight cancer. It may be an option for advanced cases of uveal melanoma.

In 2022, a new type of Immunotherapy called, Tebentafusp (Kimmtrak), was approved by the FDA to treat uveal melanoma that cannot be removed or has spread to other parts of the body. It works by bridging together tumor and immune T cells, which helps the body’s immune system to locate and kill the uveal melanoma cells. Your healthcare provider will work with you to determine the best treatment plan for your individual needs. It's important to discuss the potential benefits and risks of each treatment option, as well as any potential side effects, with your healthcare provider.

For patients facing rare melanoma subtypes, including those with uveal melanoma, it’s important to see a doctor who specializes in treating patients with your specific type of melanoma.

Is uveal melanoma curable?

No cure has been found for cancer. However, doctors consider cancer cured if you go into complete remission for 5 years. Remission means that you no longer have symptoms of the disease.

Many people who receive treatment for uveal melanoma achieve complete remission, especially if their cancer has not spread to other tissues.

The most common treatments for uveal melanoma include:

* conservative management, meaning the cancer is closely monitored without delivering any particular treatment
* radiation therapy
* laser therapies such as:
  + transpupillary thermotherapy
  + laser photocoagulation
* surgery, which might involve removing the tumor or the eye
* immunotherapy drugs like tebentafusp-tebn or darovasertib for people with metastatic cancer

outlook for people with uveal melanoma

Uveal melanoma has the best outlook when it’s diagnosed and treated before it spreads.

Doctors often use 5-year relative survival rates for reporting cancer survival. This statistic is a measure of how many people with a certain cancer are alive 5 years after receiving their diagnoses compared to people without that cancer.

The 5-year relative survival rates for eye melanoma in the United States from 2012 to 2018 were as follows:

| **Stage** | **5-year relative survival rate** |
| --- | --- |
| **Localized (no spread)** | 85% |
| **Regional (spread to nearby tissues)** | 67% |
| **Distant (spread to distant tissues)** | 16% |
| **All stages combined** | 81% |

As many as 50% of people with eye melanoma experience spread to distant organs. This is called metastatic melanoma.

About 80% of metastatic eye melanomas spread to the liver, which typically worsens the outlook. Other areas it can spread to include your:

* lungs
* skin
* soft tissues
* bone

The spread to these locations can occur 2 to 3 years after the initial diagnosis or as late as decades after.

The outlook for uvea melanoma is usually best when the tumor develops in your iris. Melanoma in the iris rarely spreads to other body parts because it’s usually detected earlier.

**DIAGNOSIS**

Diagnosis, staging and prognosis The cancer team will normally make a diagnosis based on the results of your tests and medical examination and, unlike in skin melanoma, a biopsy will usually not be necessary for diagnosis. Biopsy A biopsy removes a small amount of tissue from within the eye to examine in the laboratory. It is either done under a general anaesthetic (where you are put to sleep in hospital) or a local anaesthetic is given to numb the area. You will be able to discuss the options with your medical team. Note: If the eye is removed (enucleation), a diagnostic biopsy is done routinely, but make it clear to your surgeon if you would like a prognostic biopsy as well. Most of the time the diagnosis of uveal melanoma can be made without biopsy, but occasionally, if the diagnosis is not certain from the results of your tests and medical examination, you may be offered a biopsy to confirm the diagnosis. It will provide information about:

● Where the tumour started – although the tumour is in the eye, it may have spread from \* This may be the consultant or another senior doctor on the team. a tumour somewhere else in the body. If this is the case, treatments may be different depending on where the primary (first) tumour was located.

● What the tumour is made of which will help to confirm a diagnosis as not all intraocular (inside the eye) tumours are melanomas.

● Whether or not the tumour is cancerous. For example, it can be difficult to tell whether a small tumour is a benign mole or a melanoma without examining it under the microscope. You are likely to also be offered a biopsy to help estimate the chances of tumour spreading – a prognostic biopsy. It may help:

● Estimate the chances of the tumour spreading (metastasis) to other parts of the body. There are certain genes that increase the risk of the cancer spreading to other parts of the body and a biopsy gives an analysis of the gene profile.

● Inform future treatments - targeted therapies for tumours involving particular genes are continually being developed and knowing the genetic make-up of the tumour may be helpful in determining treatments in future. The risks of having a biopsy include:

● Minor/temporary changes in vision – there is a 10% chance that the biopsy may result in changes to your vision such as cloudy vision.

● Significant vision loss – rarely, biopsy can result in significant vision loss due to an infection, detachment of the retina, or development of a cataract.

● Very rarely, ‘seeding’ of tumour cells into tissues around the biopsy area may happen and could theoretically increase the risk of tumour spread elsewhere. ● No result – particularly if the tumour is small there is a 5–10% chance of not getting a result.

EPIDEMIOLOGY

Uveal melanoma (UM) is the most common primary intraocular malignancy in adults, with incidence rates varying significantly by geographic region, ethnicity, and gender.

Incidence Rates

* Global Range: Incidence rates generally range from about 0.4 to 10 cases per million people per year.
* By Region:
  + Europe: Incidence varies between 3.1 and 10 per million, with northern European countries like the UK (10 per million), Germany (6.4 per million), Sweden (5.6–9.6 per million), Poland (6.7 per million), and Denmark (6.5–7.8 per million) showing higher rates.
  + North America: Rates are around 5.1 to 5.2 per million.
  + Oceania: Australia reports approximately 7.6 per million, and New Zealand about 5.56 per million.
  + Asia: Much lower incidence, e.g., Republic of Korea at 0.42 per million.
  + Africa: Very low incidence, estimated at 0.2–0.3 per million.
* Gender: Incidence is slightly higher in males than females; for example, in the US, 5.8 per million in males versus 4.4 per million in females.

Trends

* The incidence of uveal melanoma has remained relatively stable over recent decades in most studied populations.
* A north-to-south gradient in Europe is observed, with higher incidence in northern countries, potentially linked to less ocular pigmentation and UV exposure differences.

Survival and Mortality

* Five-year overall survival rates range widely from about 47% to 88%, depending on the study and region.
* Ten-year survival rates are generally lower, often between 42% and 71%.
* Iris melanomas tend to have better prognosis and lower mortality compared to choroidal and ciliary body melanomas.

<https://iovs.arvojournals.org/article.aspx?articleid=2792964>

[PC\_Uveal\_Melanoma.pdf](https://melanomafocus.org/wp-content/uploads/2023/11/PC_Uveal_Melanoma.pdf)

Leber’s congenital amaurosis

Leber’s congenital amaurosis (LCA) is a rare condition that affects the retinas in babies’ eyes. Babies born with LCA have low vision — they often lose some or all of their sight. Many babies who have LCA are born blind.

LCA is a congenital condition, which means your baby is born with it. It’s caused by genetic mutations that affect how your child’s retinas develop.

The retina is the layer at the very back of your eyeball. Photoreceptors in your retinas process light into an electrical signal that your brain can understand as images you see. Rods are photoreceptors that help you see at night and in dim light. Cones process color and make up most of your usual vision.

Leber’s congenital amaurosis makes the rods and cones in your baby’s retinas malfunction. It changes how much electrical energy your child’s retinas can use and process. The less electrical activity there is, the less sight your child will have. If there’s no electrical activity in your child’s retinas, they won’t be able to see at all.

If your child has Leber’s congenital amaurosis, they’ll begin losing their sight when they’re around 6 months old. The most common treatments for LCA try to improve what vision your child does have.

Visit an eye care specialist as soon as you notice any changes in your child’s eyes or if it seems like they’re having trouble seeing.

How common is Leber’s congenital amaurosis?

Leber’s congenital amaurosis is rare. It affects around 2 out of every 100,000 babies born each year. Fewer than 50,000 people in the U.S. are living with LCA.

Even though it’s rare, LCA is one of the most common causes of blindness in children.

Symptoms and Causes

Children with Leber’s congenital amaurosis how have low vision or no vision. Because it usually develops in babies less than a year old, it might be hard or impossible for you to know something is affecting your child’s eyes.

Your child might start rubbing their eyes a lot. This is usually the first sign that something is affecting their vision. They may seem bothered by light. You might notice your child’s eyes shaking, which is called nystagmus.

Other symptoms of Leber’s congenital amaurosis include:

* Keratoconus.
* Light sensitivity (photophobia).
* Farsightedness (hyperopia).
* Slow or missing pupillary response (your child’s pupils won’t adjust to changes in light conditions).

What causes Leber’s congenital amaurosis?

Several genetic mutations can cause Leber’s congenital amaurosis. Genetic mutations are changes to your DNA that occur in your biological parent’s reproductive cells (egg and sperm) during conception.

Mutations in almost 30 different genes can cause Leber’s congenital amaurosis. The most common genetic mutations that cause LCA happen to genes that develop and form your retina, including:

* *CEP290*.
* *CRB1*.
* *GUCY2D*.
* *RPE65*.

LCA is usually an autosomal recessive condition. This means both biological parents need to pass an altered gene onto their child for their baby to inherit the genetic condition. If both parents have one of the genetic mutations that can cause LCA, there’s a 25% chance their children will develop it.

Many people carry autosomal recessive traits but don’t know it because they don’t have any symptoms. Talk to a healthcare provider about genetic counseling if you’re worried about your risk of passing genetic conditions to your children.

Diagnosis and Tests

An eye care specialist will diagnose Leber’s congenital amaurosis. They’ll perform an eye exam to look at your child’s eyes (including inside them). They’ll use electroretinography (ERG) to measure the electrical activity in your child’s retinas. Your child might need an optical coherence tomography (OCT) scan, too.

Usually, an eye care specialist will also rule out other conditions that can affect your child’s eyes. You might see this referred to as a differential diagnosis. Some conditions they’ll check for include:

* Retinitis pigmentosa.
* Joubert syndrome.
* Zellweger syndrome.
* Color blindness (achromatopsia).
* Dropping eyelids (ptosis).

Management and Treatment

There’s no cure for Leber’s congenital amaurosis. An eye care specialist will treat LCA symptoms to improve any sight your child has. Treatments to support their vision usually include eyeglasses and other low vision aids like magnifying glasses or reading prisms.

Gene therapy for Leber’s congenital amaurosis

The U.S. Food and Drug Administration (FDA) approved the first gene therapy for Leber’s congenital amaurosis in 2017. Gene therapy is an experimental treatment using genetic material to treat or prevent certain diseases. It’s currently only approved to treat LCA caused by mutations to the *RPE65* gene.

Gene therapy works by replacing or inactivating disease-causing genes. In some cases, gene therapy introduces new genes into your body to treat a specific disease. With gene therapy, doctors deliver a healthy copy of a gene to cells inside your body.

An eye care specialist will tell you if your child is a good candidate for gene therapy.

Outlook / Prognosis

You should expect your child to have little to no eyesight if they have Leber’s congenital amaurosis. Most babies born with LCA lose part or all of their vision.

Your child will need regular eye exams to track any changes in their eyes. An eye care specialist will tell you how often your child will need their eyes examined.

Prevention

You can’t prevent your child from developing Leber’s congenital amaurosis if they’ve inherited one of the genetic mutations that cause it. Talk to a provider if you’re worried about the risk of your children inheriting genetic disorders.

Living With

When should I see my healthcare provider?

Visit an eye care specialist as soon as you notice any changes in your child’s eyes or if it seems like they’re having trouble seeing.

If your child has already been diagnosed with Leber’s congenital amaurosis, see an eye care specialist if it seems like their vision or other symptoms are changing or getting worse.

**EPIDEMIOLOGY**

* Prevalence and Birth Prevalence:  
  The estimated birth prevalence of LCA is approximately 2 to 3 per 100,000 live births worldwide, which translates to roughly 1 in 33,000 to 1 in 50,000 births. This corresponds to an estimated global patient population of about 180,000 individuals.
* Proportion of Retinal Dystrophies and Childhood Blindness:  
  LCA accounts for about 5% of all inherited retinal dystrophies (IRDs) and is responsible for approximately 20% of childhood blindness cases in schools for the blind.
* Genetic and Regional Variability:  
  The prevalence of specific genetic mutations causing LCA varies by ethnicity and region. For example, mutations in *CEP290*, *GUCY2D*, and *RPE65* are more common in Caucasian populations, while *CRB1* mutations predominate in Chinese cohorts. Other mutations such as *NMNAT1* and *RPGRIP1* are more frequent in Japanese populations. These genetic differences influence the epidemiology and clinical presentation across populations.
* Incidence Trends and Risk Factors:  
  The incidence of LCA is influenced by factors such as advanced paternal age, which is associated with increased risk of inherited retinal diseases. The global birth rate (~140 million annually) suggests about 2,800 to 4,200 new LCA cases emerge each year.
* Market and Treatment Landscape:  
  The global market for LCA treatments is growing, driven by advances in gene therapy (e.g., voretigene neparvovec), though treatment costs remain a significant barrier

Differential Diagnoses

* Senior-Løken Syndrome  
  A ciliopathy combining juvenile nephronophthisis (a kidney disorder) with retinal dystrophy resembling LCA.
* Juvenile Nephronophthisis  
  A kidney disease that can be associated with retinal degeneration.
* Conorenal Syndrome (also known as Senior-Løken variant)  
  Features renal dysplasia, retinal pigmentary dystrophy, cerebellar ataxia, and skeletal abnormalities.
* Joubert Syndrome  
  Characterized by cerebellar vermis hypoplasia, early-onset retinal dystrophy, episodic hyperpnea/apnea, and developmental delay.
* Peroxisome Biogenesis Disorders  
  Includes Zellweger syndrome, neonatal adrenoleukodystrophy, and infantile Refsum disease, which present with retinal dystrophy plus systemic involvement (e.g., liver dysfunction, developmental delay).
* Infantile Neuronal Ceroid-Lipofuscinosis (CLN1)  
  Progressive neurodegenerative disorder with retinal degeneration, developmental regression, seizures, and early death.
* Mitochondrial Disorders  
  Such as Kearns-Sayre syndrome, presenting with pigmentary retinopathy, external ophthalmoplegia, and systemic features like cardiomyopathy and diabetes.
* Early-Onset Retinitis Pigmentosa (RP)  
  Has a later onset than LCA, better central vision preservation, no nystagmus, and typically shows some photopic ERG preservation, unlike the profoundly abnormal ERG in LCA.

Clinical Clues Supporting LCA

* Severe vision loss from birth or infancy
* Nystagmus present early or at birth
* Absent or severely reduced pupillary light response
* Extinguished or severely subnormal electroretinogram (ERG)
* Oculo-digital sign (eye poking/rubbing)
* Usually normal fundus in infancy, with later retinal changes

Diagnostic Approach

* Thorough clinical history and pediatric neurological examination to exclude systemic or neurodegenerative disorders
* Electroretinography (ERG) to assess retinal function
* Optical coherence tomography (OCT) for retinal structure
* Genetic testing to identify causative mutations and distinguish from overlapping inherited retinal dystrophies

REFERENCES

[Leber's Congenital Amaurosis (LCA): Symptoms & Causes](https://my.clevelandclinic.org/health/diseases/24167-lebers-congenital-amaurosis#overview)

<https://pubmed.ncbi.nlm.nih.gov/37460155/>

Leber hereditary optic neuropathy (LHON)

Leber hereditary optic neuropathy (LHON) is a genetically inherited disease that causes vision loss. Most people who inherit the condition develop blurred vision that gets progressively worse over a course of about six months. Vision loss may start in one eye and then progress to both eyes several months later. It usually starts in late childhood to early adulthood. Most people with LHON will become legally blind.

Leber hereditary optic neuropathy is also called Leber’s disease. It’s named for Dr. Theodore Leber, who studied the disease. “Hereditary” means that you inherit it, and “optic neuropathy” means it’s a disease that affects your optic nerve. Your optic nerve is what carries visual signals from your eye to your brain so that your brain can “see.” Damage to your optic nerve is one way that you can lose your vision.

Are there different types of LHON?

The standard version of Leber hereditary optic neuropathy (LHON) only affects your optic nerves, and vision loss is its only symptom. This is the case for the vast majority of people with Leber’s disease. But in rare cases, some people with LHON have additional symptoms that affect other parts of their nervous system, and sometimes their heart. Healthcare providers call this version of the disease “Leber plus.”

The true incidence of LHON is unknown. Estimates suggest that it occurs in approximately 1 in 25,000 to 50,000 people. Approximately 80% to 90% of people with LHON are male.

Symptoms and Causes

Vision loss with LHON is painless and subacute, meaning it progresses over several months. You’re likely to notice changes in your central vision first. That’s what you see when you look straight ahead — what you use to drive, read and recognize faces. Your central vision might start to seem blurry, or you might develop a blind spot in the middle of your field of vision. It might occur in one eye first, then the other.

This will continue to worsen over the following months. You also may begin to lose your color vision — the ability to see certain colors or tell colors apart. Vision loss tends to stabilize within six months to a year of the first symptoms. By this time, most people have a visual acuity of 20/200 or worse, which is legally blind. You’ll still have some light perception, but you’ll need to learn to live with low vision.

What are the symptoms of Leber’s plus?

People with Leber’s plus can have a variety of symptoms beyond vision loss, including:

* Movement disorders, like tremors.
* Coordination and balance problems (ataxia).
* Cardiac conduction problems, like arrhythmias (irregular heartbeats).
* Symptoms of multiple sclerosis (MS), like muscle weakness and fatigue.

What causes Leber hereditary optic neuropathy?

Leber hereditary optic neuropathy is a mitochondrial disease, a genetic disorder that you inherit through the mitochondria in your cells. Mitochondria are the energy generators in your cells. They convert oxygen and nutrients into the energy your cells need to operate. Mitochondrial diseases interfere with this process, leaving a lack of energy in your cells. This can cause some cells to malfunction or die.

When your mitochondria aren’t working right, the parts of your body that rely on mitochondrial energy the most will feel it first. These include parts of your eye, especially your optic nerve. If enough of your mitochondria are defective, these parts won’t have the energy they need to function right. In Leber’s disease, the effect is that your optic nerve deteriorates. This is how LHON causes vision loss.

How do you get Leber hereditary optic neuropathy?

LHON happens because of a gene mutation in your mitochondria’s DNA. Mutations in the *MT-ND1*, *MT-ND4*, *MT-ND4L* or *MT-ND6* genes can cause LHON. You inherit all of your mitochondria from your birth mother, so only female parents can pass the gene mutation to their biological children. But not everyone who carries the mutation develops symptoms of LHON, so not everyone realizes they carry it.

Risk factors might contribute to LHON

Researchers don’t know exactly why some people with the gene mutation develop symptoms of LHON and others don’t. Some evidence suggests that physiological stress and environmental toxins may contribute to triggering the onset of symptoms. The theory is that these factors may add to the overall stress on body systems affected by LHON. Over time, they add up until they finally trigger symptoms.

Potential risk factors include:

* Smoking.
* Alcohol use.
* Exposure to environmental toxins.
* Systemic illness.
* Psychological stress.

Diagnosis and Tests

Your eye care specialist will conduct standard eye exams to test your vision and look for the cause of the problem. They might not be able to see anything wrong with your eye or your optic nerve at first. The damage becomes more obvious after the first six months of symptoms. When your provider suspects LHON, they’ll recommend genetic testing to confirm that you have one of the gene mutations involved.

Management and Treatment

There’s no definitive cure for LHON. Idebenone, which is a synthetic form of Coenzyme Q10, is the only FDA-approved medication. In randomized controlled trials, idebenone has been associated with improvements in visual acuity in people with LHON. Other similar medications are currently in trials. Researchers are also studying gene therapy as a possible future treatment for Leber’s disease.

Outlook / Prognosis

When LHON causes vision loss, it’s likely to be rapid, severe and permanent. Most people will have to learn a new way of living with low vision. Low vision means moderate to severe visual impairment, but not total blindness. Moderate low vision scores 20/70 on a visual acuity test, while severe low vision scores 20/200 or worse. This is still a fairly wide range of visual acuity that you could end up with.

Whether you end up on the moderate or severe side of this range is mostly a matter of luck. Treatment might make some difference, but the gene mutation you have makes the most difference. Some people with LHON recover some of their vision unexpectedly after a year of decline. Different gene mutations have different chances of recovery. But even with some recovery, you’ll likely still have low vision.

Prevention

While many people who inherit genes associated with LHON will never develop vision loss, there’s no known way to prevent it from happening. It’s possible, but not proven, that taking antioxidants and avoiding neurotoxins like alcohol and tobacco might reduce your risk. All females who have the genes will pass them to their biological children. Genetic counseling can help you consider this risk.

**EPIDEMIOLOGY**

LHON is estimated to be the most frequent mitochondrial disease, with a prevalence ranging from 1 in 27,000 in North East England to 1 in 45,000 in a meta-analysis of reports in the European population. It has a strong male preponderance (80% to 90%), and the usual age at onset is between 15 to 35 years.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnoses for LHON include other optic neuropathies such as:

* Demyelinating optic neuritis
* Neuromyelitis optica spectrum disease
* Toxic optic neuropathy
* Compressive optic neuropathy

In addition, maculopathies and nonorganic vision loss should be considered. Clinical history, symptoms, and imagining should be utilized to differentiate LHON from these other optic neuropathies

REFERENCES

[Leber Hereditary Optic Neuropathy (LHON): Causes & Treatment](https://my.clevelandclinic.org/health/diseases/leber-hereditary-optic-neuropathy-lhon#overview)

[Leber Hereditary Optic Neuropathy (LHON) - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK482499/#article-24145.s4)

macular pucker

A macular pucker is a wrinkling of your retina due to scar tissue. Other terms for this scar tissue are epiretinal membrane (ERM) or cellophane maculopathy. The retina is the part of your eye covered with special nerve cells that react to light.

These nerve cells are very close together in the middle of your retina where the lens of your eye focuses the images that you see. The macula is the small part of your retina where the light-sensing cells come together.

A macular pucker doesn’t always cause significant issues with your vision, but it can distort your vision in some cases.

Recent studies estimate that 18.8% to 34.1% of Americans have macular pucker. The risk increases with age. Fortunately, most of these individuals don’t develop poor vision.

Symptoms and Causes

What are the symptoms of macular pucker?

The main symptom of macular pucker is distorted central vision. Straight lines appear to be wavy. Another term for this type of distorted vision is metamorphopsia.

While it can affect both eyes, it’s more common for macular pucker to affect only one eye. If you do have it in both eyes, one eye will probably be worse.

In addition to wavy vision, you might find that you aren’t able to see as clearly as you once did, no matter how close you are to what you’re looking at. You may also see a double image, or the image from one eye may seem larger than the other. Importantly, eyeglasses can’t fix the poor vision caused by macular pucker.

What causes macular pucker?

* Tears in the retina.
* Prior history of retinal detachment.
* Prior eye surgery.
* Eye trauma.
* Infection.
* Diabetes-related retina disease.
* Eye inflammation.

Is macular pucker contagious?

No, macular pucker isn’t contagious. You can’t get it from anyone or spread it to anyone else.

Risk factors for macular pucker

In addition to getting older, you may be at a higher risk of macular pucker if you:

* Have a separation of the vitreous humor from the retina.
* Had laser eye therapy or eye surgery, including cataract surgery.
* Had a retinal tear or detachment.
* Had uveitis or eye inflammation.
* Have diabetes-related retinopathy.
* Had severe eye injury.

Complications of macular pucker

Macular pucker may lead to some vision loss. It isn’t likely to cause blindness.

Diagnosis and Tests

How is macular pucker diagnosed?

Your eye care provider will do a thorough eye exam after asking you about your symptoms and taking a medical history. They’ll give you eye drops to dilate your eyes (make them open wide) so they can see your retina.

What tests will be done to diagnose macular pucker?

During your eye exam, your provider will use a light and magnifier to get a close look at your eyes. They may also use the following tests to diagnose macular pucker:

* Amsler grid eye test, which checks for distorted vision using a page of small squares formed by horizontal and vertical lines.
* Optical coherence tomography, an imaging method that takes pictures of your retina.

Management and Treatment

You may not need treatment for macular pucker. Your provider may consider just monitoring your condition. If you do need treatment, there are noninvasive and surgical options.

Noninvasive treatments for macular pucker

* A new prescription and new eyeglasses. Eyeglasses won’t fix the problems caused by macular pucker, but a new prescription may be able to optimize and improve your overall vision.
* Good lighting, such as reading lamps.
* Magnifying devices.

Surgical treatments for macular pucker

* Vitrectomy with membranectomy, an outpatient procedure that removes the scar tissue or membrane from the retina.

What are the complications of treating macular pucker with surgery?

Your healthcare team will take every precaution to avoid complications, but there’s always a small risk during any surgery, including:

* Bleeding.
* Infection.
* Retinal tear or detachment.
* Worsening of cataracts.
* Macular hole.
* Glaucoma or high eye pressure.

How soon after treatment will I feel better?

After a vitrectomy, you’ll probably have imperfect vision for a few days. Your eyes may be irritated and tender for a little while.

You’ll probably need to take two to four weeks off work or school.

You won’t be able to drive right away. It could even take up to 3 months to enjoy the full benefits of improved vision after surgery.

Outlook / Prognosis

Most cases of macular pucker don’t need treatment. You’ll just need to have eye exams on the schedule your provider sets up for you to monitor your vision and eye health.

You may have some vision problems with macular pucker, but it’s not likely to cause blindness.

Can macular pucker be prevented?

In some cases, where a cause is unknown, there’s no way of preventing it. Avoiding diabetes-related eye disease and eye trauma are some ways to help prevent it.

Living With

How do I take care of myself if I have a macular pucker?

Ways to take care of yourself include:

* Keeping your scheduled eye appointments.
* Following general guidelines for good overall health, like eating healthy foods and getting appropriate amounts of exercise.
* Following guidelines for good eye health, like wearing sunglasses, washing your hands before touching your eyes and following your provider’s advice about wearing contact lenses safely.
* Stop smoking. Smoking isn’t good for your eyes. If you smoke, ask your healthcare provider for help with quitting.

When should I see my healthcare provider?

If you have any changes in your eyesight, you should contact your healthcare provider. If you have sudden changes in vision or sudden eye pain, get immediate medical help by calling 911 or going to the emergency room

Epidemiology

Frequency

United States

The frequency at which epiretinal membranes occur varies according to the underlying disease. The idiopathic variety of epiretinal membranes has been shown to be present in up to 7% of the population. Bilateral cases have been seen in as much as 30% of the population. A 2016 examination of the Beaver Dam Eye Study cohort using OCT suggested a higher prevalence of epiretinal membrane in the population (34.1%).

Clinically significant epiretinal membranes occur in 3-8.5% of eyes after successful primary retinal detachment surgery. Patients noted to be at the greatest risk for epiretinal membranes are those with preoperative signs of proliferative vitreoretinopathy, including rolled retinal edges, star folds, and equatorial ridges.

One study noted no significant difference in the frequency of epiretinal membranes in eyes that underwent retinal detachment repair with subretinal fluid drainage compared to those that had nondrainage procedures. While there is the belief that excessive cryotherapy or laser photocoagulation may lead to increased risk of ERM formation, the possible risk of epimacular development in eyes that have undergone cryotherapy or laser photocoagulation for retinal tears is difficult to quantify because it is almost impossible to determine whether the cellular dispersion was caused by the retinal tear itself or the subsequent therapy for it..

Differential Diagnoses of Epiretinal Membrane (ERM)

Epiretinal membrane (ERM), also known as macular pucker or cellophane maculopathy, is characterized by a fibrocellular membrane on the inner retinal surface, often affecting the macula and causing visual distortion. Although the clinical appearance of ERM is distinctive, several other conditions can mimic or be confused with ERM. The main differential diagnoses include:

* Macular Hole  
  A full-thickness defect in the macula that can appear similar to an ERM pseudohole but differs in retinal layer involvement. Macular holes often cause more profound vision loss and require different management.
* Parafoveal Telangiectasia (Macular Telangiectasia Type 2)  
  A vascular disorder of the macula that can cause retinal thickening, cystic changes, and subtle surface changes, sometimes resembling ERM on clinical exam.
* Macular Edema  
  Swelling of the macula due to fluid accumulation from various causes (e.g., diabetic retinopathy, retinal vein occlusion) can cause retinal thickening and visual distortion, potentially confused with ERM.
* Retinal Vascular Diseases  
  Conditions such as diabetic retinopathy or retinal vein occlusion can lead to secondary ERM formation and must be differentiated.
* Uveitis and Other Inflammatory Diseases  
  Chronic inflammation can cause vitreoretinal interface changes and secondary ERM.
* Retinal Tears or Detachments  
  Peripheral retinal pathology can coexist or mimic ERM symptoms and should be ruled out with thorough examination.
* Intraocular Tumors  
  Rarely, tumors can cause secondary ERM formation; fluorescein angiography and imaging help differentiate.
* Pseudohole  
  A gap or hole in the ERM itself that mimics a macular hole but without full-thickness retinal defect.

REFERENCES

[Epiretinal Membrane Differential Diagnoses](https://emedicine.medscape.com/article/1223882-differential?form=fpf)

[Macular Pucker: What It Is, Symptoms & Treatment](https://my.clevelandclinic.org/health/diseases/14207-macular-pucker)

macular hole

A macular hole is an actual hole or full-thickness defect in the macula of your eye, the central part of your retina. Your retina is the part of your eye that contains nerve cells that react to light and allow you to see. A macular hole generally happens in just one eye, but it can happen in both eyes.

In the middle of the retina (the macula), the nerve cells are very close together. Sometimes, the jelly-like substance that fills your eye — the vitreous humor — changes its consistency and, as it shrinks, it can pull on the central macula and cause a macular hole to form. A macular hole can affect your vision in a variety of ways, but it mainly affects your central vision (the things that you can see in the center of your visual field). It affects activities like driving and reading.

How common is this condition?

One U.S. study indicates that the number of new cases of macular hole is 7.8 people per 100,000 people per year. It's more common in women. It happens much more often in people aged 55 and over, but it can happen to anyone.

Are there types of macular holes?

There are primary and secondary macular holes. A primary macular hole is one that develops without any eye injury and isn’t due to another medical condition. A secondary macular hole is one that occurs with or due to another disease or condition, such as trauma or eye inflammation (uveitis).

There are a couple of ways that ophthalmologists may classify macular holes. These are called staging systems. One system uses four stages, with the first stage being the least severe and the fourth being the most severe. A newer system has three stages based on the results of imaging tests. Your eye care provider will give you information on how severe your macular hole is.

Symptoms and Causes

A macular hole is a gap in the macula, which is the center of the retina of your eye.

What are the symptoms of a macular hole?

The early symptoms of a macular hole include:

* Blurred vision.
* Distorted vision. Straight lines might be curvy or wavy.
* Difficulty reading small print.

A later sign of a macular hole is a dark or blind spot in the center of your vision.

If any of these symptoms occur, it’s important to make an appointment with your eye care provider as soon as possible.

What causes a macular hole?

In most people, macular holes are due to vitreous traction that’s more likely to happen with aging. Sometimes a macular hole is the result of an injury or a medical condition that affects the eye, including being very nearsighted.

You may be more likely to develop a macular hole if you have:

* A very high degree of myopia (nearsightedness).
* Inflammation within your eye (uveitis).
* Eye trauma (injury).

Risk factors for developing a macular hole

Risk factors for macular holes include:

* Aging.
* Female sex.
* History of eye trauma.
* Being very nearsighted.
* Previous eye surgeries or eye inflammation.

What are the complications of a macular hole?

An untreated macular hole may lead to these complications:

* A loss of vision, especially central vision.
* A macular-hole-associated ret1``inal detachment.
* Increased size of the macular hole.

Diagnosis and Tests

Your eye care provider will begin by asking you about your family and medical history. You’ll want to tell them about any type of medication — prescription and over-the-counter (OTC) drugs — that you take. Your provider will then do a complete eye exam, which will include a slit lamp exam. Your provider will put eye drops in your eyes to make your pupils larger and allow for retina examination.

What tests will be done to diagnose a macular hole?

Your ophthalmologist will probably order one or more of these tests to examine your retinas:

* Optical coherence tomography.
* Fundus fluorescence angiography (also called fluorescein angiography).
* Fundus photography.

Management and Treatment

How is a macular hole treated?

The most common treatment for macular holes is a procedure called a vitrectomy. A vitrectomy is a surgery during which a retina specialist removes the vitreous gel of your eye. Your surgeon, an ophthalmologist trained in retina surgery, may also remove any bits of tissue (membranes) that may be putting tension on your macula.

Your surgeon will put a sterile gas into your eye to keep pressure on the hole until it heals. You may have to stay in a facedown position for one to seven days to keep the bubble in place so the hole will close.

If you have a small hole, your retina specialist may suggest watching and waiting rather than treating it. Sometimes an early-stage macular hole will close on its own.

What are the complications of a vitrectomy for a macular hole?

Possible complications of a vitrectomy may include:

* Retinal detachment.
* Infection.
* Glaucoma.
* Bleeding.
* The macular hole reopening or never closing.

Cataracts are expected to progress in any person who’s having retina surgery.

How soon after treatment will I feel better?

If you have a vitrectomy, you may need to spend up to a week with your head down to keep the gas bubble in the correct position. The gas bubble lasts three to eight weeks, and your vision remains blurry while the gas bubble’s in your eye. It may take several months for your vision to improve and stabilize.

Outlook / Prognosis

What can I expect if I have a macular hole?

The success rate for vitrectomy surgeries is over 90%. The surgery is most successful when the hole is smaller and more recent. You may regain most or some of your lost vision.

When can I go back to work or school?

You’ll need to take some time away from work or school. The length of time may vary depending on the type of work you do. Ask your surgeon about what you need to do or to avoid doing after the procedure.

You probably won’t be able to drive for six to eight weeks, and you won’t be able to fly for about that long, either. It takes this long for your body to absorb the gas. Changes in air pressure may make the bubble in your eye get bigger and dangerously increase the pressure in the eye. Avoid any type of activity that involves changes in air pressure.

What is the outlook for a macular hole?

If you get treatment sooner, or if the hole is small, your prognosis (outlook) is good.

Without treatment, you may lose much of your central vision, but you’ll retain your peripheral (side) vision.

Prevention

Can a macular hole be prevented?

There’s no way to prevent a macular hole. You may reduce your risk by:

* Using protective eye coverings when working or participating in contact sports.
* Having regular eye examinations. This may not prevent a macular hole, but your provider will be able to find a macular hole earlier if you follow a recommended exam schedule.
* Taking care of your eyes and managing your blood sugar levels, if you have diabetes, and your blood pressure, if you have hypertension.

Living With

When should I see my healthcare provider?

Everyone should get regular eye appointments. Apart from these, see your eye care provider any time you notice a change in your vision. If you experience extreme pain, or if you have a sudden loss of vision, go to an emergency room.

Epidemiology

Frequency

United States

The overall prevalence is approximately 3.3 cases in 1000 in those persons older than 55 years. Peak incidence of idiopathic macular hole development is in the seventh decade of life, and women typically are affected more than men. Reasons for this, at best, are speculative at this point. Some epidemiologic risk factors, such as cardiovascular disease, hypertension, and a history of hysterectomy, have been reported by other studies. However, none of these have been proven to have any significant association with macular hole formation.

International

The prevalence rate of macular hole in India is a reported 0.17%, with a mean age of 67 years.

The Beijing Eye Study found the rate of macular holes to be 1.6 out of 1000 elderly Chinese, with a strong female predilection.

Mortality/Morbidity

The natural history of a macular hole varies based on its current clinical stage. It has been reported that around 50% of stage 0 and stage 1 macular holes may resolve both in the anatomic changes and the symptoms produced. Stage 2 holes progress and worsen in most cases to stage 3 or stage 4, resulting in worsening vision. Best estimates for the incidence of development of an idiopathic full-thickness macular hole in the fellow eye are approximately 12%.

Differential Diagnosis of Macular Hole (MH)

Macular hole, a full-thickness defect in the neurosensory retina at the fovea, must be differentiated from several other vitreomacular interface and macular conditions that can mimic its clinical appearance or symptoms.

Key Differential Diagnoses

* Epiretinal Membrane (ERM) with Pseudohole  
  ERM can cause a macular pseudohole, which appears as a round, well-defined opening but without a full-thickness retinal defect. Unlike a true macular hole, the retinal layers remain intact, and OCT shows no full-thickness break.
* Lamellar Macular Hole (LMH)  
  A partial-thickness defect involving the inner retinal layers but sparing the outer retina. LMH shows irregular foveal contour with a break in the inner fovea on OCT but no full-thickness retinal defect.
* Vitreomacular Traction (VMT) Syndrome  
  Characterized by persistent vitreous adhesion causing distortion of the foveal contour and intraretinal changes without a full-thickness hole. VMT can progress to MH but is distinct on OCT.
* Cystoid Macular Edema (CME)  
  Fluid accumulation within the macula causing cystic spaces and retinal thickening that may mimic early or impending macular hole on clinical exam.
* Central Foveal Dot Hemorrhage  
  Small hemorrhages at the fovea can cause visual symptoms and mimic early MH lesions.
* Subfoveal Drusen and Adult Vitelliform Macular Dystrophy  
  These can cause central macular changes and visual distortion resembling early MH or stage 1 MH.
* Other Conditions to Consider
  + Early stage 1 macular hole (impending hole) with loss of foveal depression but no full-thickness defect.
  + Traumatic macular hole or secondary MH associated with high myopia, diabetic retinopathy, or other retinal diseases.

[Macular Hole Differential Diagnoses](https://emedicine.medscape.com/article/1224320-differential?form=fpf)

[Macular Hole: Symptoms, Causes, & Treatment](https://my.clevelandclinic.org/health/diseases/14208-macular-hole#overview)

Macular Edema

Even as the word retina has become commonplace, the macula and its diseases are often misunderstood. The retina is the light-sensitive layer of cells that lines the inside of the eye.

The many layers of the retina work together to convert light focused on the retina into an exquisitely detailed message that travels to the visual cortex in the brain. There, the message is decoded and directs us to take action— “that’s a fine looking piece of pie!”

The macula is the part of the retina that helps us see fine detail, faraway objects, and color. It’s packed with more photoreceptors (light-sensitive cells) than any TV or monitor. The small, central area of the retina is worth the most—the bullseye of sight. Macular edema, degeneration, hole, pucker, drusen (small yellowish deposits), scar, fibrosis, hemorrhage, and vitreomacular traction are common conditions that involve the macula. When macular disease is present, distorted vision (metamorphopsia), blank spots (scotoma), and blurred vision are common symptoms.

Risk factors

Macular edema is not a disease, but is the result of one.

As with other conditions where abnormal fluid accumulates (leg swelling, pulmonary edema, hives, and allergy), macular edema can be caused by many factors including

* Metabolic conditions (diabetes)
* Blood vessel diseases (vein occlusion/blockage)
* Aging (macular degeneration)
* Hereditary diseases (retinitis pigmentosa)
* Traction on the macula (macular hole, macular pucker, and vitreomacular traction)
* Inflammatory conditions (sarcoidosis, uveitis)
* Toxicity
* Neoplastic conditions (eye tumors)
* Trauma
* Surgical causes (following eye surgery)
* Unknown (idiopathic) causes

Macular edema occurs when the retina’s ability to absorb fluid is overwhelmed by the fluid leaking into it. If more rain falls on the lawn than it can handle, you get puddles of water. In the retina, blisters of fluid form and swell the retina—this is macular edema. Factors likely to cause macular edema include conditions that:

* Cause more fluid to leak from blood vessels (diabetes and high blood pressure)
* Increase inflammation in the eye (surgery, inflammatory diseases)
* Are associated with the growth of abnormal blood vessels (wet age-related macular degeneration)

Symptoms

Macular edema refers to an abnormal blister of fluid in the layers of the macula. From the side, it looks like the snake that ate too much. Like a droplet of water on your computer screen, the swollen retina distorts images—making it more difficult to see clearly. The more widespread, thicker, and severe the swelling becomes, the more likely one will notice visual symptoms of blur, distortion, and difficulty reading.

If untreated, chronic macular edema can lead to irreversible damage of the macula and permanent vision loss. Macular edema is typically caused by increased leakage from damaged retinal blood vessels or growth of abnormal blood vessels in the deep retina. New vessels (neovascularization or NV) do not have normal “tight junctions” and almost always result in abnormal leakage of fluid (serum from the bloodstream) into the retina.

Treatment and prognosis

The most effective treatment strategies for macular edema address the underlying cause (diabetes, blood vessel occlusion, neovascularization, inflammation, etc), as well as an excess of fluid leaking from abnormal blood vessels in and around the macula. Eye drops, laser, and surgery can be effective in many diseases, but the mainstay of treatment is intravitreal injections (IVI).

The IVI is an office procedure performed under topical anesthesia in which medication is placed inside the eye by a very small needle. The injection genrally causes little to no pain. IVI should be performed by a trained retina specialist with meticulous monitoring of treatment efficacy and detection of rare but potentially serious complications. IVI is now considered one of the most commonly performed medical procedures.

Ranibizumab (Lucentis®), aflibercept (Eylea®), and dexamethasone (Ozurdex®) are the generic and trade names, respectively, of the 3 most widely used FDA-approved medications for IVI treatment of the common conditions causing macular edema.

Bevacizumab (Avastin®) is not FDA approved for this use, but has also been extensively studied in large, well-designed, federally funded clinical trials and is felt to have excellent efficacy. US physicians are permitted to use drugs in a manner not included in the FDA’s approved packaging label; this common practice is known as off-label use.

Each treatment option has a considerable track record of success and works by decreasing the amount of fluid leaking from abnormal blood vessels. There are differences between each of these drugs. Your retina specialist will work with you to identify which options are best for you.

Macular edema is a common finding in many diseases of the retina, almost all of which can be treated to improve vision. As with other conditions, the visual prognosis depends on the severity of the underlying condition, its duration, the general health of the eye, and the degree to which vision has been affected.

There has never been a more successful time in the treatment of macular edema from a variety of causes, and more promising therapies will be available in the future.

causes macular edema?

Macular edema occurs when fluid or blood leaks into the retina. This builds up and causes swelling, resulting in changes in a person’s vision.

The cells of the retina line the back of the eye, sensing light and sending signals to the brain. This is how the eye creates images. The macula sits at the center of the retina, allowing the eye to see color, distant objects, and small details in the visual field.

Macular edema occurs when this area of the retina swells. However, macular edema is not an eye condition on its own. Instead, it usually develops due to conditions that cause fluid to leak into the back of the eye.

Diagnose macular edema

If an ophthalmologist, a type of eye doctor, suspects macular edema, they may use eye drops to expand the pupils and examine the eye.

The doctor may then perform various tests to help confirm the diagnosis.

Fluorescein angiography

Fluorescein angiography involves injecting a yellow dye into a vein in the arm. This then travels through the blood vessels, including those at the back of the eye.

The ophthalmologist can use a specialized camera to create images of the retina as the dye travels through it, showing the location and extent of any blood vessel leakage.

Optical coherence tomography (OCT)

OCT uses highly detailed cameras. The resulting images show the thickness of the macula, allowing an ophthalmologist to see how swollen it is and where leakage may be occurring.

Amsler grid

The ophthalmologist may also use an Amsler grid to identify problems with the center of a person’s visual field.

An ophthalmologist shows the individual a grid of lines. If part of it looks wavy, they may investigate central vision loss, which is a symptom of macular edema.

Epidemiology of Macular Edema (Primarily Diabetic Macular Edema)

* Global Prevalence:  
  Diabetic macular edema (DME) affects approximately 5.5% to 6.8% of people with diabetes worldwide, translating to about 19 to 29 million individuals globally. The prevalence is projected to increase as the global diabetic population grows, with estimates rising from 19 million currently to 29 million by 2045.
* Prevalence by Region and Income Level:  
  The pooled global prevalence of DME diagnosed by optical coherence tomography (OCT) is about 5.47%, with a similar prevalence in low-to-middle-income countries (5.81%) and high-income countries (5.14%). Regional studies show variability, with prevalence in Africa ranging higher (e.g., up to 20.8% in South Africa and 33.3% in Kenya) compared to Western countries where prevalence ranges from about 3.8% to 11.1%.
* Prevalence in Specific Countries:
  + United States: Approximately 3.8% prevalence among diabetic adults aged 40 and older, equating to roughly 746,000 individuals.
  + Ethiopia: Hospital-based studies report a prevalence of 17% for DME and 5.5% for clinically significant macular edema (CSME).
  + Western societies generally report prevalence between 3.8% and 7.12%.
* Risk Factors Associated with DME:  
  Major risk factors include longer duration of diabetes, poor glycemic control (elevated HbA1c), systemic hypertension, proteinuria, dyslipidemia, insulin therapy, and history of cataract surgery.
* Trends:  
  The prevalence of diagnosed DME has increased over recent years, with a reported 1.5-fold increase in annual diagnosis from 2.8% in 2009 to 4.3% in 2018 in some populations.
* Burden:  
  DME is the leading cause of vision loss among people with diabetes and is a major contributor to diabetic retinopathy-related visual impairment

**DIFFERENTIAL DIAGNOSIS**

Macular edema can be caused by several other conditions including but not limited to hypertension, retinal vein occlusion, ruptured microaneurysm, radiation, Irvine-Gass syndrome, and subfoveal choroidal neovascularisation. A good clinical history can help to elucidate the underlying cause of the macular edema. Questions about the patient's onset of diabetes, hemoglobin A1C trending, diet, and presence of other diabetic complications are critical

REFERENCES

[Diabetic Macular Edema - StatPearls - NCBI Bookshelf](https://www.ncbi.nlm.nih.gov/books/NBK554384/#article-24631.s9)

Proliferative Vitreoretinopathy

Proliferative vitreoretinopathy is a condition where abnormal cells grow on the retina, causing scarring and potentially leading to vision loss. This disease can significantly impact the individual's quality of life by affecting their vision and overall well-being. It can result in severe visual impairment and even blindness, affecting daily activities and independence. Early detection and appropriate management are essential to prevent further vision deterioration and improve the patient's health outcomes.

**Types of Proliferative Vitreoretinopathy**

Proliferative vitreoretinopathy (PVR) can manifest in various forms, each with distinct characteristics and implications for vision. These different types of PVR may affect the retina and vitreous humor differently, leading to diverse clinical presentations and treatment approaches. Understanding the specific type of PVR a patient has is essential for devising an appropriate management plan and achieving optimal visual outcomes.

Epiretinal Membrane (ERM): A thin layer of fibrous tissue that forms on the surface of the retina, leading to visual distortion or blurriness.

Subretinal Proliferative Vitreoretinopathy: Abnormal growth of cells beneath the retina, causing detachment and vision problems.

Retrolental Membrane: Formation of scar tissue behind the lens of the eye, impacting vision and potentially leading to retinal detachment.

Circumferential Proliferative Vitreoretinopathy: Proliferation of cells around the circumference of the retina, resulting in traction and distortion of the retina.

Posterior Proliferative Vitreoretinopathy: Development of scar tissue at the back of the eye, affecting the retina's function and structure, often leading to vision loss.

**Risk Factors**

Proliferative vitreoretinopathy (PVR) is a condition that can affect the eyes, particularly after certain types of eye surgeries or trauma. There are several factors that can increase the risk of developing PVR. Understanding these risk factors is essential for managing the condition effectively.

Previous eye surgeries

Severe eye trauma

Retinal detachment

Inflammatory eye conditions

Prolonged retinal detachment

High myopia

Diabetes

Intraocular hemorrhage

Causes of Proliferative Vitreoretinopathy

Proliferative vitreoretinopathy (PVR) is a complication that can occur after eye surgery or trauma. It is mainly caused by the growth of abnormal cells on the retina's surface leading to scar tissue formation. Other factors such as inflammation, retinal detachment, and certain eye conditions can also contribute to the development of PVR. Early detection and prompt treatment are crucial in managing this condition effectively.

* Retinal detachment
* Trauma to the eye
* Intraocular surgery
* Prolonged inflammation
* Proliferative diabetic retinopathy

Symptoms of Proliferative Vitreoretinopathy

Proliferative vitreoretinopathy (PVR) is a condition that affects the retina and the vitreous in the eye. It is characterized by certain symptoms that may impact vision and overall eye health. Symptoms can vary in severity and may include changes in vision, discomfort, and other noticeable signs. Early detection and prompt treatment are essential in managing PVR and preventing potential complications. Regular eye exams are crucial for individuals at risk of developing this condition.

* Blurred vision
* Floaters in vision
* Flashes of light
* Distorted vision
* Reduced visual acuity
* Vision loss

Diagnosis of Proliferative Vitreoretinopathy

Proliferative vitreoretinopathy is diagnosed through various eye exams and tests conducted by an eye specialist. These tests help determine the severity of the condition and guide treatment decisions. Early diagnosis is crucial to prevent vision loss and effectively manage the disease. By identifying the signs and symptoms, healthcare providers can recommend appropriate interventions to improve the patient's vision and overall eye health.

* Ophthalmoscopy
* Optical coherence tomography (OCT)
* Ultrasound imaging
* Fluorescein angiography
* B Scan ultrasonography

Treatment for Proliferative Vitreoretinopathy

Proliferative vitreoretinopathy (PVR) is a condition that can lead to vision loss if left untreated. Treatment options for PVR aim to prevent further progression of the disease and preserve vision.

* Surgical Removal of Scar Tissue: In cases of proliferative vitreoretinopathy, surgery may be necessary to remove the abnormal scar tissue that is pulling on the retina and causing vision problems.
* Vitrectomy: A vitrectomy is a surgical procedure that involves removing the vitreous gel from the eye. This can help improve vision by relieving traction on the retina caused by proliferative vitreoretinopathy.
* Intravitreal Injections: Medications such as steroids or antiVEGF agents can be injected into the eye to help reduce inflammation and abnormal blood vessel growth associated with proliferative vitreoretinopathy.
* Retinal Detachment Repair: Repairing a detached retina is often necessary in cases of proliferative vitreoretinopathy to restore vision and prevent further complications.
* Silicone Oil Tamponade: In some cases, a silicone oil bubble may be used to help stabilize the retina after surgery for proliferative vitreoretinopathy, promoting healing and preventing further detachment.

## **Epidemiology of Proliferative Vitreoretinopathy (PVR)**

* Incidence in Retinal Detachment:  
  Proliferative vitreoretinopathy occurs in approximately 5–10% of all cases of rhegmatogenous retinal detachment (RRD), making it the most common cause of failure after retinal detachment surgery. Despite advances in surgical techniques, this incidence has remained relatively stable over recent decades.
* Prevalence in Retinal Detachment Cases:  
  Among patients with retinal detachment, the prevalence of any type of PVR can be as high as 52.9%, with severe PVR occurring in about 26.9% of cases, often associated with longer duration of detachment and delayed surgery.
* Incidence after Ocular Trauma:  
  PVR is much more frequent following ocular trauma, especially open-globe injuries, where it develops in 40–60% of cases. The incidence varies by type of injury, with rates of approximately 43% after perforation, 21% after rupture, 15% after penetration, 11% with intraocular foreign bodies, and 1% after contusion injuries.
* Risk Factors:  
  Factors increasing PVR risk include prolonged retinal detachment duration, delayed surgical intervention, ocular trauma, presence of vitreous hemorrhage, and intraocular inflammation.
* Recurrent Retinal Detachment:  
  PVR is the leading cause of recurrent retinal detachment, with most recurrences occurring within three months post-surgery.
* Market and Treatment Trends:  
  The global market for PVR treatment and management is growing, with an estimated value nearing US$660 million by 2027 and a compound annual growth rate (CAGR) of about 4%

## **Differential Diagnoses**

* Proliferative Vascular Retinopathies
  + Proliferative diabetic retinopathy (PDR)
  + Other ischemic proliferative retinopathies
  + Retinopathy of prematurity (ROP)  
    These conditions involve neovascular proliferation and fibrous tissue formation but are primarily vascular in origin, unlike PVR which follows retinal detachment.
* Retinal Detachment Following Ocular Trauma
  + Retinal detachment secondary to open-globe injuries or intraocular foreign bodies can lead to retinal folds and scarring that mimic PVR.
  + Trauma-related detachments may have associated inflammation and hemorrhage complicating the picture.
* Rhegmatogenous Retinal Detachment (RRD) Without PVR
  + Some cases of RRD, especially in highly myopic eyes with thin retinas, may show extensive retinal folds that can appear fixed but actually retain mobility on eye movement, differentiating them from PVR.
  + These cases lack the contractile membranes typical of PVR and the retina usually flattens completely after surgery.
* Other Fibrosing or Scar-Forming Conditions
  + Inflammatory or infectious retinopathies that cause vitreoretinal fibrosis may mimic aspects of PVR.
  + Conditions causing vitreous opacities and tractional membranes should be differentiated by history, clinical exam, and imaging.

REFERENCES.

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