

## Uveitis

Uveitis refers to a rare group of inflammatory diseases of the uveal tissue in the eye. The uvea includes the choroid, ciliary body and the iris. The uvea is rich in blood vessels and is the source of the inflammatory cells that enter the eye. Uveitis inflammation can damage the lens, retina, optic nerve or the vitreous. Uveitis is estimated to be responsible for approximately 10%-20% of the blindness in the United States. Uveitis treatment is challenging since it can damage many parts of the eye. It can cause minor symptoms or lead to severe vision loss. Uveitis affects approximately 1 in 4500 people and is most common between the ages 20 to 60 with men and women affected equally. It can also occur in children and seniors. Uveitis may be caused by problems or diseases isolated to the eye or it can be part of an inflammatory disease affecting other parts of the body. It may have slow or sudden onset and last for days to many years with recurrences. Uveitis may develop suddenly with redness and pain or with just a blurring of vision.

## Uveitis Classification

Uveitis is classified based on the cause as infectious or non-infectious (autoimmune) or by the area of the eye involved as

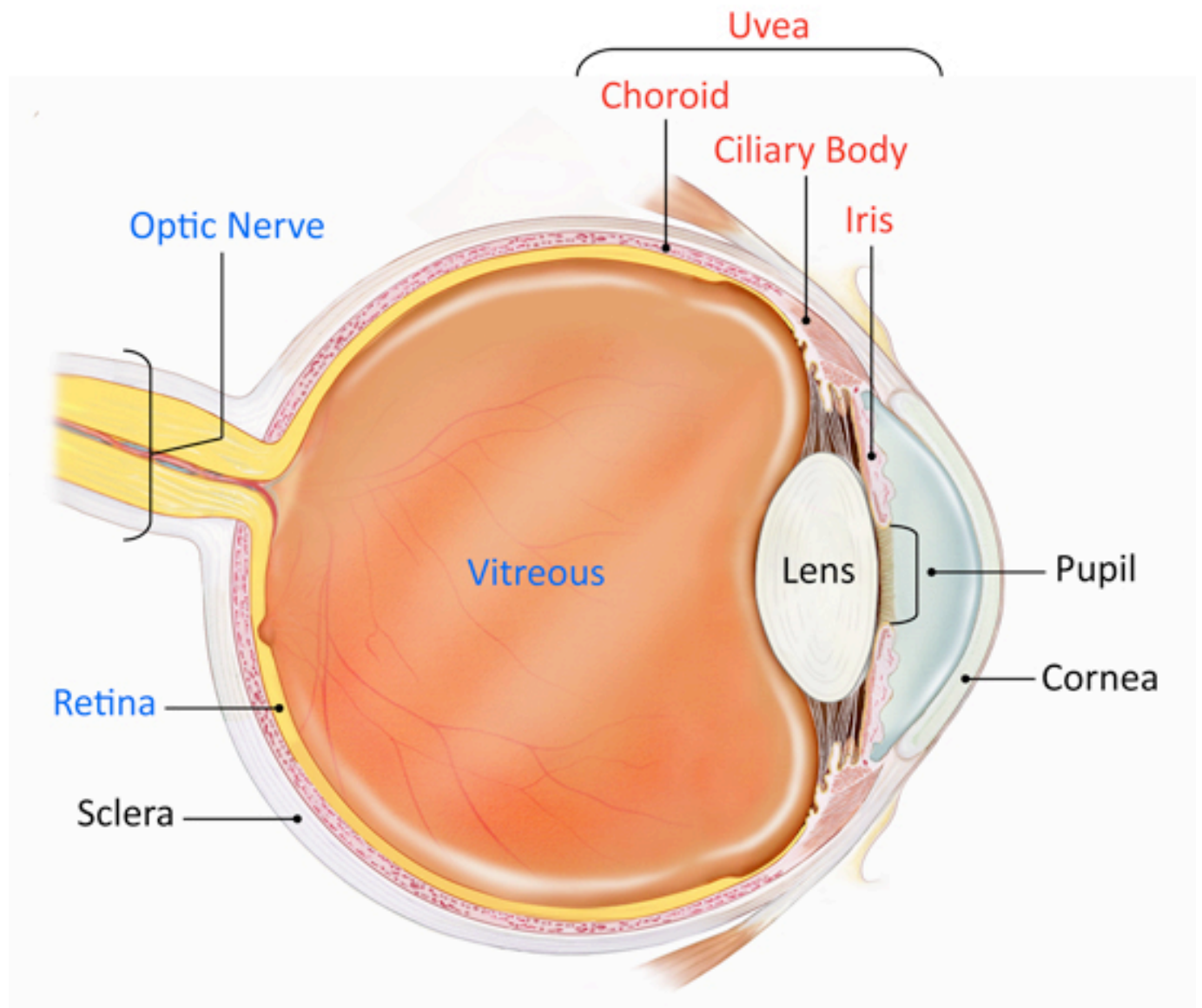
- **Anterior uveitis:** Anterior uveitis occurs in the front of the eye. It is the most common form of uveitis, predominantly occurring in young and middle-aged people. Many cases occur in healthy people and may only affect one eye but some are associated with joint, skin, gastrointestinal, lung and infectious diseases.
- **Intermediate uveitis:** Intermediate uveitis is commonly seen in young adults. The center of the inflammation often appears in the vitreous. It has been linked to several disorders including, sarcoidosis and multiple sclerosis.
- **Posterior uveitis:** Posterior uveitis is the least common form of uveitis. It primarily occurs in the back of the eye, often involving both the retina and the choroid. It is often called choroditis or chorioretinitis. There are many infectious and non-infectious causes to posterior uveitis.
- **Panuveitis uveitis:** This can affect all parts of the eye

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The uveal tissue includes the Choroid, ciliary body and the iris.

Lens: Transparent tissue that allows light into the eye.

*Retina*: The layer of cells on the back, inside part of the eye that converts light into electrical signals sent to the brain.

*Optic Nerve*: A bundle of nerve fibers that transmits electrical signals from the retina to the brain.

*Vitreous*: The fluid filled space inside the eye.

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## What are the Symptoms of Uveitis?

Uveitis can affect one or both eyes. Symptoms may develop gradually or rapidly and are variable in severity and may last weeks to years and include:

- Blurred vision or a change in vision
- Dark, floating spots in the vision (floaters)
- Eye pain or burning of the eye
- Redness of the eye ringing the eye or throughout the white portion
- An irregular shaped pupil
- Light sensitivity or photophobia
- Headaches

## How is Uveitis Diagnosed?

Diagnosis of uveitis includes obtaining an extensive medical history and performing a thorough eye examination with dilation of the pupils and a neurologic exam in some cases. Laboratory tests may be done to rule out an infection or an autoimmune disorder and x-rays may be taken of the spine and chest. If a specific cause or infection is suspected, more specific treatment can be utilized.

## Uveitis Classification

Uveitis is classified based on the cause as infectious or non-infectious (autoimmune) or by the area of the eye involved as:

- **Anterior uveitis:** Anterior uveitis occurs in the front of the eye. It is the most common form of uveitis, predominantly occurring in young and middle-aged people. Many cases occur in healthy people and may only affect one eye but some are associated with joint, skin, gastrointestinal, lung and infectious diseases. In anterior uveitis, no associated condition or syndrome is found in approximately one-half of cases. However, anterior uveitis is often one of the syndromes associated with HLA-B27.
- **Intermediate uveitis:** Intermediate uveitis is commonly seen in young adults. The center of the inflammation often appears in the vitreous. It has been linked to several disorders including, sarcoidosis and multiple sclerosis. It is also called pars planitis.
- **Posterior uveitis:** Posterior uveitis is the least common form of uveitis. It primarily occurs in the back of the eye, often involving both the retina and the choroid. It is often called choroditis or chorioretinitis. There are many infectious and non-infectious causes to posterior uveitis.
- **Panuveitis uveitis:** Pan-uveitis is when all three major parts of the eye are affected by inflammation. Behcet's disease is one of the most well-known forms of pan-uveitis and it greatly damages the retina.

Intermediate, posterior, and panuveitis are the most severe and highly recurrent forms of uveitis. They often cause blindness if left untreated.

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## What Causes Uveitis?

The causes of uveitis may be classified as:

- **Infectious**
  - **AIDS**
  - brucellosis
  - CMV
  - herpes zoster
  - leptospirosis
  - Lyme disease
  - presumed ocular histoplasmosis syndrome
  - syphilis
  - toxocariasis
  - toxoplasmic chorioretinitis
  - tuberculosis
  - Vaccinations
  - Zika fever[9]
  
- **Noninfectious or autoimmune causes**
  - Behçet disease
  - Crohn's disease
  - Fuchs heterochromic iridocyclitis
  - Granulomatosis with polyangiitis
  - HLA-B27 related uveitis
  - Juvenile idiopathic arthritis
  - Kawasaki disease
  - Multiple sclerosis
  - polyarteritis nodosa
  - Sarcoidosis
  - Spondyloarthritis
  - Sympathetic ophthalmia following eye injury
  - Tubulointerstitial nephritis and uveitis syndrome
  - Ulcerative colitis
  - Vogt Koyanagi Harada's disease
  - Whipple Disease

- **Associated with systemic diseases.**

In most cases, uveitis is an isolated illness only involving the eyes but there is no association of uveitis with other diseases. However, uveitis can be associated with many diseases including:

- AIDS
- Ankylosing spondylitis
- Behcet's syndrome
- CMV retinitis
- Eye injuries
- Herpes zoster infection
- Histoplasmosis
- Kawasaki disease

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- Multiple sclerosis
- Psoriasis
- Reactive arthritis
- Rheumatoid arthritis
- Sarcoidosis
- Syphilis
- Toxoplasmosis
- Tuberculosis
- Ulcerative colitis
- Vogt Koyanagi Harada's disease
- **Drug related side effects**
  - [Rifabutin](#)
  - quinolone antibiotics such as [Moxifloxacin](#)
  - all widely administered vaccines
- **White Dot syndromes**
  - [acute posterior multifocal placoid pigment epitheliopathy](#)
  - [birdshot chorioretinopathy](#)
  - [multifocal choroiditis and panuveitis](#)
  - [multiple evanescent white dot syndrome](#)
  - [punctate inner choroiditis](#)
  - [serpiginous choroiditis](#)
  - [acute zonal occult outer retinopathy](#)

## What are Masquerade Syndromes?

Masquerade syndromes may also mimic uveitis with intraocular cells resembling inflammatory cells within the eye but are not due to immune-mediated uveitis entities. These may represent cancer (neoplastic) involving the eye or non-neoplastic conditions.

- **Non-neoplastic:**
  - [retinitis pigmentosa](#)
  - intraocular foreign body
  - [juvenile xanthogranuloma](#)
  - [retinal detachment](#)
- **Neoplastic:**
  - [retinoblastoma](#)
  - [lymphoma](#)
  - [malignant melanoma](#)
  - [leukemia](#)
  - [reticulum cell sarcoma](#)

## How is Uveitis Treated?

Because uveitis is a serious condition that can cause permanent damage to the eye, it needs to be treated as soon as possible. Uveitis treatments primarily try to eliminate inflammation, alleviate pain, prevent further tissue damage, and restore any loss of vision. Treatment may be very specific or generalized depending on the type of uveitis a patient displays. If an infectious etiology is found, it is specifically treated. In non-infectious cases, drops to dilate the pupil and steroids are usually initiated in the form of eye drops, injections around or inside the eye. Steroids

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may also be administered by mouth but oral or intravenous immunosuppressive medications may be used when the disease is occurring in both eyes, particularly in the back of both eyes that may be safer than long-term steroid therapy that can produce side effects such as stomach ulcers, osteoporosis (bone thinning), diabetes, cataracts, glaucoma, cardiovascular disease, weight gain, fluid retention, and Cushing's syndrome. Usually other agents are started if it appears that patients need moderate or high doses of oral steroids for more than 3 months.

Other immunosuppressive agents that are commonly used include medications such as methotrexate, mycophenolate, azathioprine, and cyclosporine. These treatments require regular blood tests to monitor for possible side effects. In some cases, biologic response modifiers (BRM), or biologics, such as, adalimumab, infliximab, daclizumab, abatacept, and rituximab are used. These drugs target specific elements of the immune system. Some of these drugs may increase the risk of having cancer. If uveitis is associated with other conditions like glaucoma or retinal damage, surgery may be required

## **Prognosis**

The prognosis is generally good for those who receive prompt diagnosis and treatment, but serious complication include:

- cataracts
- glaucoma
- band keratopathy
- cystoid macular edema
- permanent vision loss may result if left untreated.

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