

Uveal Tract (Vascular pigmented layer)

Basic Knowledge

- **Structure:** The uveal tract (also known as the vascular pigmented layer, vascular tunic, and uvea)

The uveal tract consists of the following structures:

1. Iris,
2. Ciliary body,
3. Choroid.

Position: The uveal tract lies between the sclera and retina.

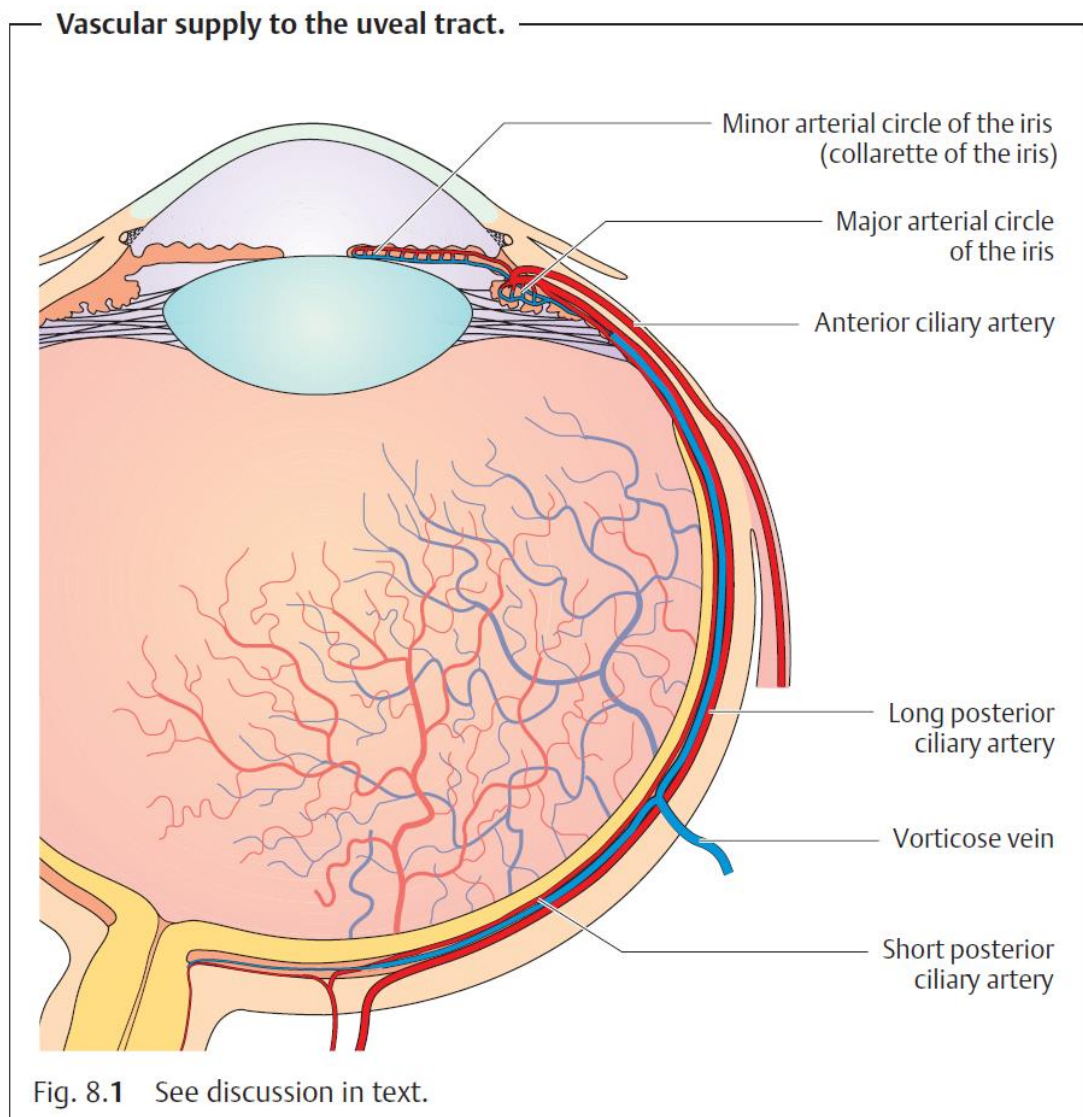
Neurovascular supply: *Arterial supply* to the uveal tract is provided by the **ophthalmic artery**.

- The **short posterior ciliary arteries** enter the eyeball with the optic nerve and supply the **choroid**.
- The **long posterior ciliary arteries** course along the interior surface of the sclera to the **ciliary body** and the **iris**. They form the major arterial circle at the root of the iris and the minor arterial circle in the **collarette** of the iris.
- The **anterior ciliary arteries** originate from the vessels of the rectus muscles and communicate with the **posterior ciliary vessels**.
- Venous blood drains through four to eight vorticoses or vortex veins that penetrate the sclera posterior to the equator and join the superior and inferior ophthalmic veins (Fig. 8.1)
- Sensory supply is provided by the long and short ciliary nerves.

Iris

Structure and function: The iris consists of two layers:

1. The anterior mesodermal stromal layer.
 2. The posterior ectodermal pigmented epithelial layer.
- The posterior layer is opaque and protects the eye against excessive incident light .
 - The anterior surface of the lens and the pigmented layer are so close together near the pupil that they can easily form adhesions in inflammation.



- The **collarette of the iris** covering the minor arterial circle of the iris divides the stroma into **pupillary** and **ciliary** portions.
- The pupillary portion contains the **sphincter muscle**, which is supplied by parasympathetic nerve fibers, and the **dilator pupillae muscle**, supplied by sympathetic nerve fibers.
- These muscles regulate the contraction and dilation of the pupil so that the iris may be regarded as the **aperture** of the optical system of the eye.
- Pupil dilation is sometimes sluggish in preterm infants and the newborn because the dilator pupillae muscle develops relatively late.

Surface: The normal iris has a richly textured surface structure with **crypts** (tissue gaps) and interlinked **trabeculae**. A faded surface structure *can* be a Sign of inflammation (iritocyclitis).

Color: The color of the iris varies in the individual according to the **melanin content of the melanocytes (pigment cells)** in the *stroma* and *epithelial layer*.

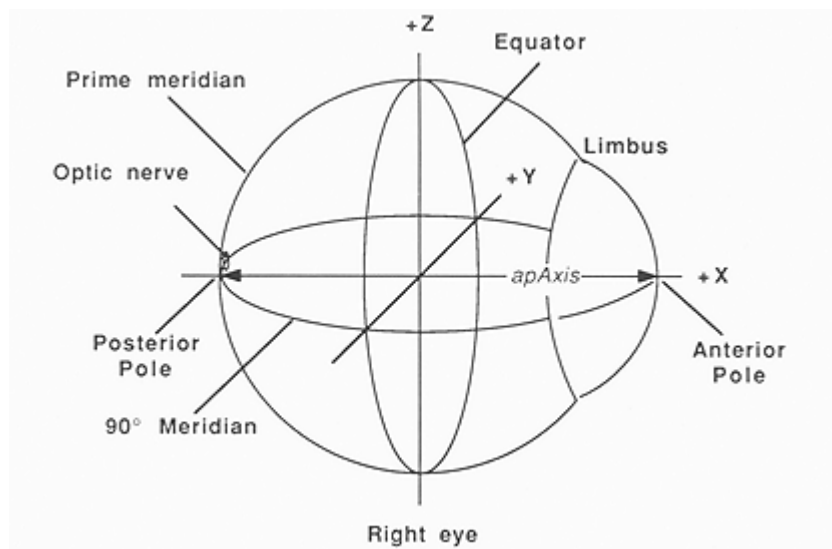
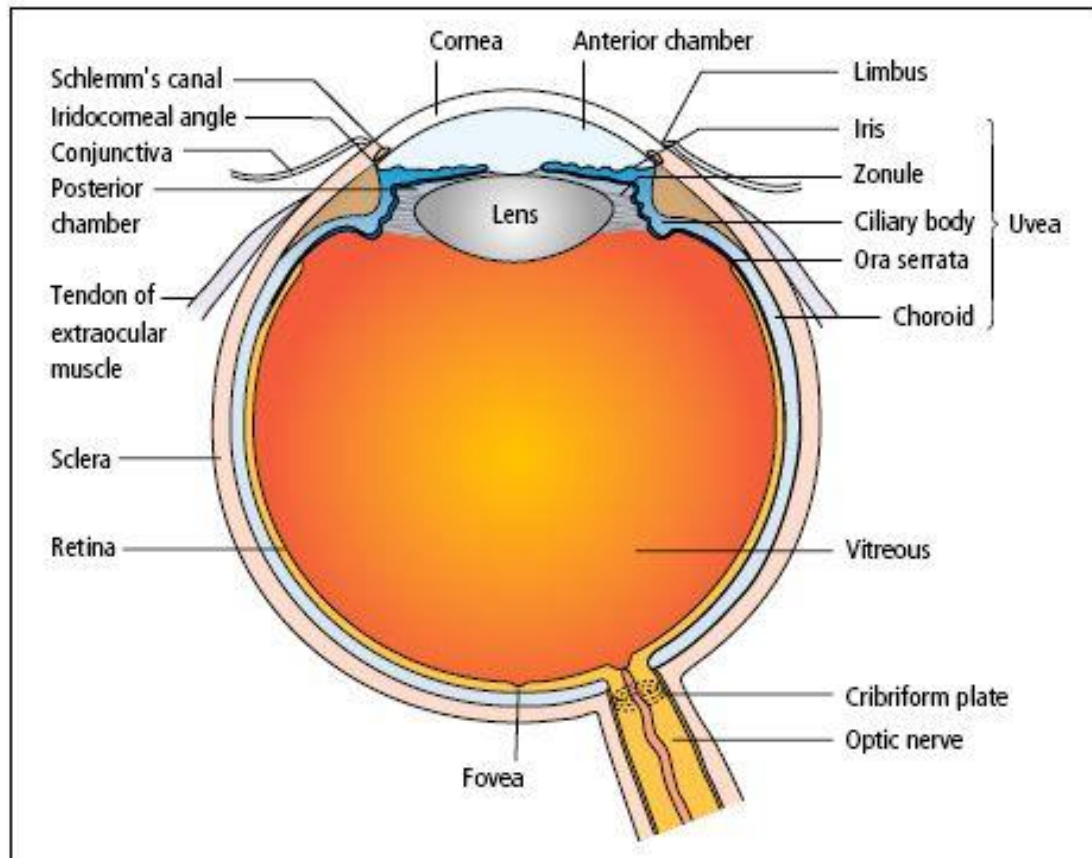
- Eyes with a high melanin content are dark brown, whereas eyes with less melanin are grayish-blue. **Caucasians at birth** always have a grayish blue iris as the *pigmented layer* only develops gradually during the first year of life.
- Even in **albinos**, the eyes have a **grayish blue iris** because of the melanin deficiency. Under slit lamp retroillumination, they appear reddish due to the fundus reflex.

Ciliary Body

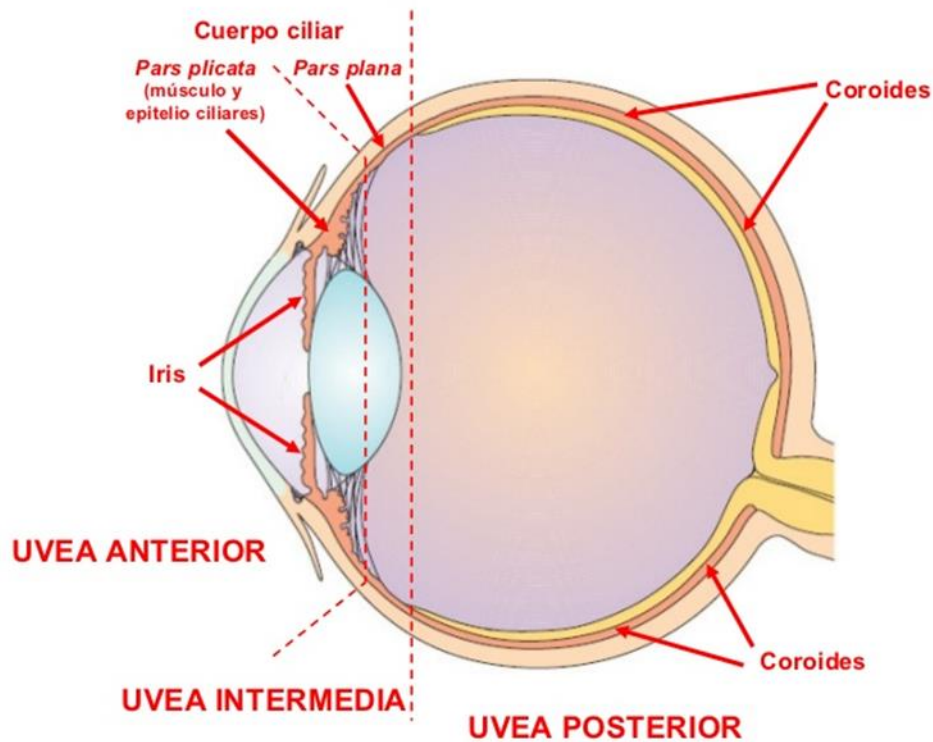
Position and structure: The **ciliary body** extends from the root of the iris to the ora serrata, where it joins the choroid. It consists of **anterior pars plicata** and the **posterior pars plana**, which lies 3.5mm posterior to the limbus.

Numerous **ciliary processes** extend into the posterior chamber of the eye. The suspensory ligament, the zonule, extends from the pars plana and the intervals between the ciliary processes to the lens capsule.

Function: The *ciliary muscle* is responsible for **accommodation**. The double layered *epithelium covering the ciliary body produces the aqueous humor*.



What is the equator of the eye? It is an imaginary line encircling the globe of the eye equidistant from the anterior and posterior poles.(see the picture above)



Choroid

Position and structure: The choroid is the **middle tunic of the eyeball**. It is bounded on the interior by **Bruch's membrane**.

- The choroid is highly vascularized, containing a vessel layer with large blood vessels and a capillary layer.
- The blood flow through the choroid is the *highest in the entire body*.

Function: The choroid **regulates temperature** and supplies **nourishment to the outer layers of the retina**.

Examination Methods

- The **slit lamp** is used to examine the **surface of the iris** under a focused beam of light. *Normally no vessels will be visible.*
- Iris vessels are only visible in atrophy of the iris, inflammation, or as neovascularization in **rubeosis iridis** (see Fig. 8.12).
- Where vessels are present, they can be visualized by *iris angiography* after intravenous injection of fluorescein sodium dye.

Neovascularization in the iris: rubeosis iridis.

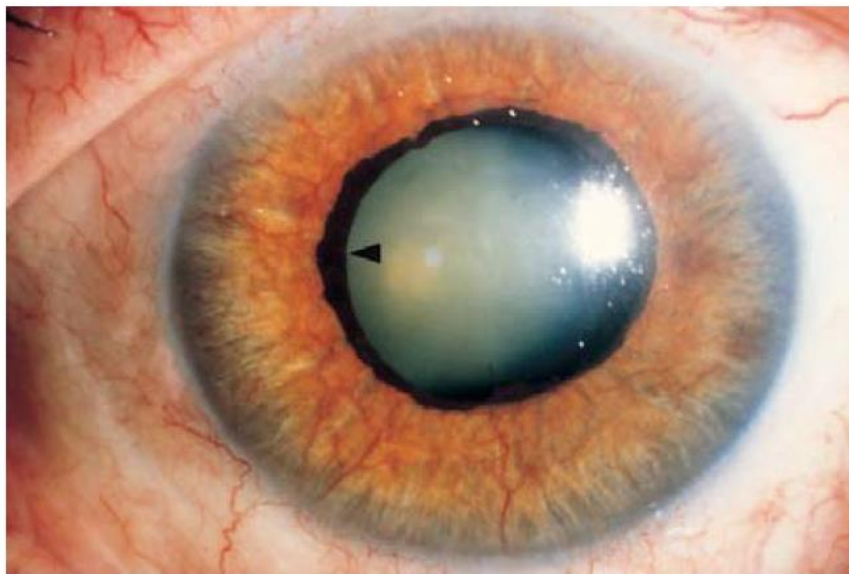


Fig. 8.12 Protrusion of the pigmented layer (arrow) indicates that the rubeosis iridis has been present for at least several weeks.

- **Defects in the pigmented layer of the iris** appear red under retro-illumination with a slit lamp (see Fig. 8.6).
- **Slit lamp biomicroscopy** visualizes individual cells such as melanin cells at 40-power magnification.
- The *anterior chamber* is normally transparent.
- Inflammation can increase the permeability of the **vessels of the iris** and compromise the barrier between blood and aqueous humor.
- **Opacification of the aqueous humor** by proteins may be observed with the aid of a *slit lamp* when the eye is illuminated with a lateral focal beam of light (Tyndall effect).

Ocular albinism.

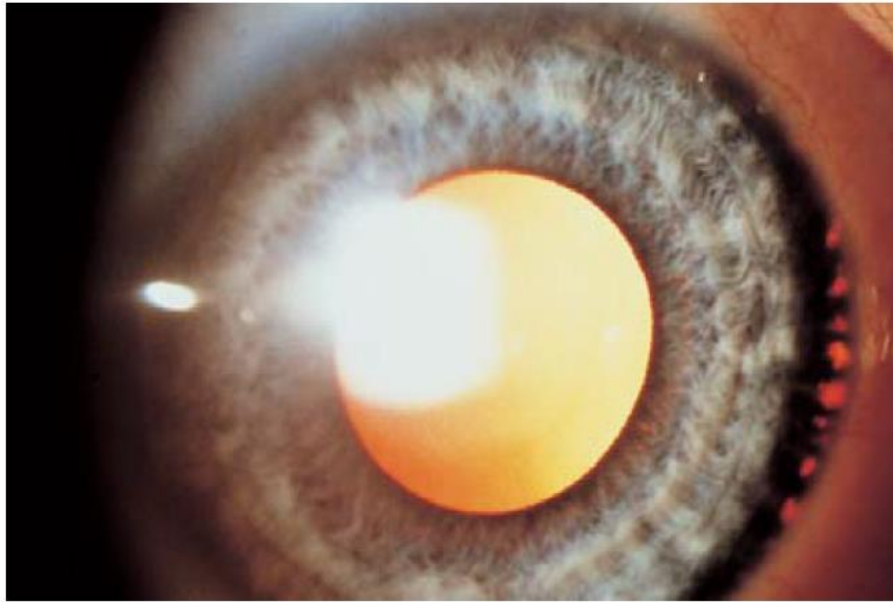


Fig. 8.6 The peripheral iris appears red under retroillumination.

- Changes in the choroid such as tumors or hemangiomas can be visualized by ultrasound examination.
- Where a **tumor is suspected**, transillumination of the eye is indicated.
- After administration of topical anesthesia, a fiberoptic light source is placed on the eyeball to *visualize the shadow of the tumor on the red of the fundus*.

Inflammation

Inflammations of the uveal tract are classified according to the various portions of the globe:

- Anterior uveitis (**iritis**).
- Intermediate uveitis (**cyclitis**).
- Posterior uveitis (**choroiditis**).

However, some inflammations involve the middle portions of the uveal tract such as **iridocyclitis** (inflammation of the iris and ciliary body) or **panuveitis** (inflammation involving all segments).

Definition of Uveitis: it is an inflammation of the uveal tract (Iris, Ciliary body and choroid) and adjacent structures, most probably the retina.

Classification: -

- Anatomical.
- Clinical.
- Aetiological.

Anatomical Classification:

1- Anterior uveitis: which is subdivided into:

a- Iritis: in which inflammation predominantly affects the iris.

b- Iridocyclitis: in which both the iris and anterior part of the ciliary body (pars plicata) are equally involved.

If the number inflammatory cells was equal in both the aqueous and vitreous, it is iridocyclitis, while if the number was larger in aqueous, it is iritis.

2- Intermediate uveitis:

It is characterized by involvement predominantly of the posterior part of the ciliary body (pars plana), periphery of the retina and the underlying periphery of the choroid.

3- Posterior uveitis:

Inflammation of the choroid and retina posterior to the equator of the eye.

4- Panuveitis:

Involvement of the entire uveal tract.

Clinical classification:

1- Acute uveitis: usually has a sudden, symptomatic onset and persists for up to 3 months. If the inflammation recurs following the initial attack it is referred as recurrent acute uveitis.

2- Chronic uveitis: the onset is frequently insidious and *may be* asymptomatic. It usually persists for longer than 3 months. Acute or subacute exacerbations on chronic may occur.

Aetiological classification:

1. **Idiopathic:** which forms more than 50% of cases of uveitis.

2. **Associated with a systemic disease, e.g.:**

a) Spondyloarthropathies: ankylosing spondylitis, Reiter's syndrome, psoriatic arthritis and chronic juvenile arthritis.

b) Inflammatory bowel disease: ulcerative colitis, Crohn's disease, Whipple's disease.

c) Nephritis.

d) Non-infectious multi-system disease: sarcoidosis, Behçet's disease.

e) Infectious systemic disease: e.g. TB, syphilis

f) Diabetes.

3. **Infections:**

a) Bacterial: tuberculosis.

b) Fungal: Candidiasis.

c) Viral: Herpes Zoster.

4. **Infestations:**

a) Protozoa: Toxoplasmosis.

b) Nematodes: Toxocariasis.

Acute Iritis and Iridocyclitis

Epidemiology: Iritis is the most frequent form of uveitis. It usually occurs in combination with cyclitis. About three-quarters of all iridocyclitis cases have an acute clinical course.

Etiology: Iridocyclitis is frequently attributable to **immunologic causes** such as allergic or hyperergic reaction to bacterial toxins. In some rheumatic disorders it is known to be frequently associated with the expression of specific human leukocyte antigens (HLA) such as HLA-B27. Iridocyclitis can also be a **symptom of systemic disease** such as ankylosing spondylitis, Reiter's syndrome, sarcoidosis, etc. (Table 8.1). **Infections** are less frequent and occur secondary to penetrating trauma or sepsis (bacteria, viruses, mycosis, or parasites).

Table 8.1 Causes of uveitis according to location

Form of uveitis	Possible causes
HLA-B27-associated iridocyclitis	<ul style="list-style-type: none"> ❖ Idiopathic ❖ Ankylosing spondylitis ❖ Reiter's syndrome ❖ Regional enteritis ❖ Ulcerative colitis ❖ Psoriasis
Non-HLA-B27-associated iridocyclitis	<ul style="list-style-type: none"> ❖ Idiopathic ❖ Viral ❖ Tuberculosis ❖ Sarcoidosis ❖ Syphilis ❖ Leprosy ❖ Rheumatoid arthritis (Still-Chauffard syndrome) ❖ Heterochromic cyclitis ❖ Phacogenic uveitis ❖ Trauma

Iridocyclitis and choroiditis

- ❖ Toxoplasmosis
- ❖ Sarcoidosis
- ❖ Tuberculosis
- ❖ Syphilis
- ❖ Behçet's disease
- ❖ Sympathetic ophthalmia
- ❖ Borreliosis
- ❖ Brucellosis
- ❖ Yersiniosis
- ❖ Listeriosis
- ❖ Malignant tumors

Choroiditis

- ❖ Toxoplasmosis
- ❖ Sarcoidosis
- ❖ Syphilis
- ❖ Behçet's disease
- ❖ Histoplasmosis
- ❖ Toxocara

Phacogenic inflammation, possibly with glaucoma, can result when the lens becomes involved

Symptoms: Patients report dull pain in the eye or forehead accompanied by impaired vision, photophobia, and excessive tearing (epiphora).

- In contrast to choroiditis, acute iritis or iridocyclitis is painful because of the involvement of the ciliary nerves.

Diagnostic considerations: Typical signs include:

- **Ciliary injection:** The episcleral and perilimbal vessels may appear blue and red.
- **Combined injection:** The conjunctiva is also affected.
- The **iris** is **hyperemic** (the iris vessels will be visible in a light-colored iris).
- The **structure** appears **diffuse** and **reactive miosis** is present.
- Vision is impaired because of cellular infiltration of the anterior chamber and protein or fibrin accumulation (visible as a **Tyndall effect**). The precipitates accumulate on the posterior surface of the cornea in a triangular configuration known as

Arlt's triangle. Exudate accumulation on the floor of the anterior chamber is referred to as **hypopyon** (Fig. 8.8). Viral infections may be accompanied by bleeding into the anterior chamber (**hyphema**; Fig. 8.9).

- Corneal edema can also develop in rare cases.

Hypopyon in acute iridocyclitis.



Fig. 8.8 The purulent exudate accumulates as a pool on the floor of the anterior chamber.

Hyphema.



Fig. 8.9 Bleeding into the anterior chamber can occur in rubeosis iridis, trauma, or, in rare cases, iridocyclitis.

- Corneal edemas and Tyndall effects (accumulations of protein in the anterior chamber) can be diagnosed when the eye is illuminated with a lateral beam of light from a focused light or slit lamp.

Differential diagnosis: See Table 8.2.

- In acute iritis, the depth of the anterior chamber is normal and reactive miosis is present. In contrast, in acute glaucoma the anterior chamber is shallow and the pupil is dilated (Table 8.2).

Table 8.2 Differential diagnosis of iritis and acute glaucoma

Differential criteria	Acute iritis	Acute glaucoma
Symptoms	Dull pain and photophobia	Intense pain and vomiting
Conjunctiva	Combined injection	Combined injection
Cornea	Clear	Opacified, edematous
Anterior chamber	Normal depth; cells and fibrin are present	Shallow
Pupil	Narrowed (reactive miosis)	Dilated, not round
Globe	Normal pressure	Rock hard

Complications: These include:

1. **Secondary open angle glaucoma** with an increase in intraocular pressure.
2. Adhesions between the iris and posterior surface of the cornea (**anterior synechiae**).
3. Adhesions between the iris and lens (**posterior synechiae**; Fig. 8.10).

Posterior synechiae secondary to iridocyclitis (cloverleaf pupil).

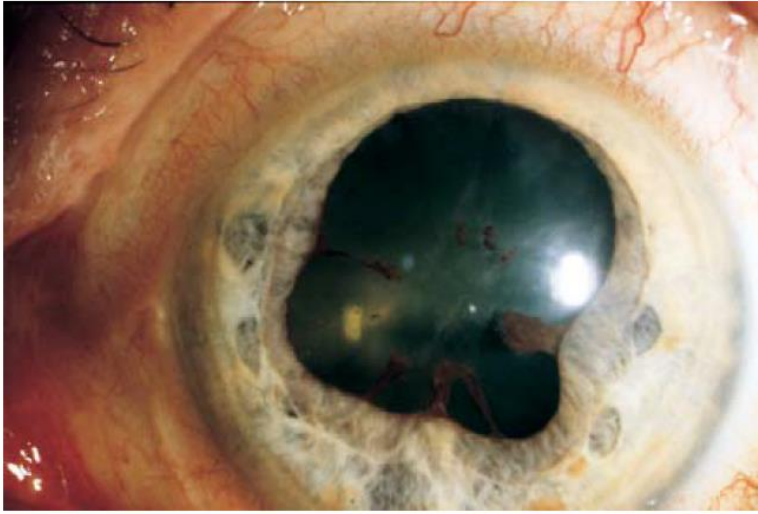


Fig. 8.10 Acute iridocyclitis produces adhesions between the iris and lens (see also Fig. 8.5).

Treatment:

- Topical and, in appropriate cases, systemic antibiotic or antiviral therapy is indicated for iridocyclitis due to a pathogen (with a corneal ulcer, penetrating trauma, or sepsis).
- A conjunctival smear, or a blood culture in septic cases, is obtained to identify the pathogen.
- Antibiotic therapy should begin immediately as microbiological identification of the pathogen is not always successful.
- Therapeutic mydriasis in combination with steroid therapy is indicated to **minimize the risk of synechiae**.
- Where no pathogen can be identified, high-dose topical steroid therapy (prednisolone eyedrops every hour in combination with subconjunctival injections of soluble dexamethasone) is administered.
- To minimize the risk of *posterior synechiae*, the pupil must be maximally dilated (atropine, scopolamine, cyclopentolate, and possibly epinephrine and epinephrine eyedrops).
- The mydriatic effect of dilating eyedrops may be reduced in iritis. This may necessitate the use of longer-acting medications such as atropine, which may have to be applied several times daily.

- **Secondary open angle glaucoma** is treated by administering beta blockers in eyedrop form and, in applicable cases, carbonic anhydrase inhibitors (acetazolamide; see Table 10.3).

Prognosis: Symptoms usually improve within a few days when proper therapy is initiated. The disorder can progress to a chronic stage.

Chronic Iritis and Iridocyclitis

Epidemiology: About one quarter of all **iridocyclitis** cases have a chronic clinical course.

Etiology: See Table 8.1.

Symptoms: See acute iridocyclitis. Chronic iridocyclitis may exhibit minimal symptoms.

Diagnostic considerations: See acute iridocyclitis.

Differential diagnosis: The disorder should be distinguished from acute glaucoma, conjunctivitis, and keratitis.

Complications:

1. Total obliteration of the pupil by posterior synechiae is referred to a **pupillary block**.
2. Because the aqueous humor can no longer circulate, **secondary angle closure glaucoma with iris bombé** occurs.
3. **Occlusion of the pupil** also results in fibrous scarring in the pupil. This can lead to the development of posterior subcapsular opacities in the lens (**secondary cataract**).
4. Recurrent iridocyclitis can also lead to calcific band keratopathy.

Treatment: In **pupillary block** with a secondary angle closure glaucoma, a *Nd:YAG laser iridotomy* may be performed to create a shunt to allow the aqueous humor from the posterior chamber to circulate into the anterior chamber. In the presence of a **secondary cataract**, a *cataract extraction* may be performed when the inflammation has abated.

Prognosis: Because of the chronic recurrent course of the disorder, it frequently involves complications such as synechiae or cataract that may progress to blindness from shrinkage of the eyeball.

Choroiditis

Epidemiology: There are few epidemiologic studies of choroiditis. The annual incidence is assumed to be four cases per 100 000 people.

Etiology: See Table 8.1.

Symptoms:

- Patients are free of pain, although they report blurred vision and floaters.
- Choroiditis is painless as the choroid is devoid of sensory nerve fibers.

Diagnostic considerations: Ophthalmoscopy reveals isolated or multiple choroiditis foci. In *acute disease* they appear as ill-defined white dots (Fig. 8.11). *Once scarring has occurred* the foci are sharply demarcated with a yellowish-brown color. Occasionally the major choroidal vessels will be visible through the atrophic scars.

Multifocal choroiditis.



Fig. 8.11 The foci of acute inflammation are yellowish and ill-defined; older lesions are yellowish-brown and sharply demarcated.

- *No cells will be found in the vitreous body in a primary choroidal process.*
- However, inflammation proceeding from the retina (**retinochoroiditis**) will exhibit *cellular infiltration of the vitreous body.*

Differential diagnosis: This disorder should be distinguished from retinal inflammations, which are accompanied by cellular infiltration of the vitreous body and are most frequently caused by viruses or *Toxoplasma gondii*.

Treatment: Choroiditis is treated either with antibiotics or steroids, depending on its etiology.

Prognosis: The inflammatory foci will heal within two to six weeks and form chorioretinal scars. The scars will result in localized scotomas that will reduce visual acuity if the macula is affected.

Sympathetic Ophthalmia

Definition

Specific bilateral inflammation of the uveal tract due to chronic irritation of one eye, caused by a perforating wound to the eye or intraocular surgery, produces transferred uveitis in the fellow eye.

Epidemiology: Sympathetic ophthalmia is very rare.

Etiology: Sympathetic uveitis can occur in an *otherwise unaffected* eye even years after penetrating injuries or intraocular surgery in the fellow eye, especially where there was chronic irritation. Tissues in the injured eye (uveal tract, lens, and retina) act as antigens and provoke an autoimmune disorder in the unaffected eye.

Symptoms: The earliest symptoms include limited range of accommodation and photophobia. Later there is diminished visual acuity and pain.

Diagnostic considerations: Clinical symptoms include combined injections, cells and protein in the anterior chamber and vitreous body, papillary and retinal edema, and granulomatous inflammation of the choroid.

Differential diagnosis: The disorder should be distinguished from iridocyclitis and choroiditis from other causes (see Table 8.1).

Treatment:

- The injured eye, which is usually blind, must be enucleated to eliminate the antigen.
- High-dose topical and systemic steroid therapy is indicated.
- Concurrent treatment with immunosuppressives (cyclophosphamide and azathioprine) may be necessary.
- Clinical course and complications: The disorder has a chronic clinical course and may involve severe complications of uveitis such as secondary glaucoma, secondary cataract, retinal detachment, and shrinkage of the eyeball.
- Sympathetic ophthalmia can lead to blindness in particularly severe cases.
- When the injured eye is blind, prophylactic enucleation is indicated before the onset of sympathetic ophthalmia in the fellow eye.
- An early sign of sympathetic ophthalmia is a limited range of accommodation with photophobia

Tumors

Malignant Tumors (Uveal Melanoma)

- With an incidence of one per ten thousand, malignant uveal melanoma is the most common primary intraocular tumor. It usually occurs as a choroidal melanoma, and is almost always unilateral. *Tumors in the iris* are detected earlier than tumors located in the *ciliary body* and *choroid* (Fig. 8.13).
1. **Iris melanomas:** These tumors are *often initially asymptomatic*. However, metastatic melanoma cells in the angle of the anterior chamber can lead to *secondary glaucoma*. Circumscribed iris melanomas are removed by *segmental iridectomy*.

Choroidal melanoma.



Fig. 8.13 A prominent yellowish-brown choroidal tumor (thick arrowheads) accompanied by serous retinal detachment (arrows).

2. Ciliary body melanomas:

- Symptoms include changes in accommodation and refraction resulting from displacement of the lens. Ciliary body melanomas are resected *en bloc*.

3. Choroidal melanomas: These tumors become clinically symptomatic when involvement of the macula *reduces visual acuity* or the patient notices a shadow in his or her field of vision as a result of the tumor and the accompanying *retinal detachment*. The diagnosis is confirmed with the aid of transillumination, ultrasound, and fluorescein angiography.

- Choroidal tumors are treated with radioactive isotopes delivered by plaques of radioactive material (*brachytherapy*).
- *Enucleation* is indicated for tumors whose diameter exceeds 8mm and whose prominence exceeds 5mm.

4. Uveal metastases most frequently develop from carcinomas of the breast or lung. They are usually flat with little pigmentation.

Benign Choroidal Tumors

- Choroidal nevi occur in 11% of the population. They can lead to secondary neovascularization with retinal edema. In very rare cases where the macula is involved, choroidal nevi can lead to impaired vision. However, benign choroidal tumors are normally asymptomatic.

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