UVEA / UVEITIS

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Uvea = middle layer (coat) of the globe
      = vascular pigmented layer

- A. iris
- B. ciliary body
- C. choroid

- Position: the uveal tract lies between the sclera and retina.
- Arterial supply to the uveal tract is provided by the ophthalmic artery.
Iris

- It is a coloured, circular diaphragm with a central aperture — the pupil, measuring about 3.5 - 4 mm. It regulates the amount of light rays reaching the retina.

- Iris consists of 2 layers:
  - **Anterior stromal layer**
  - **Posterior pigmented epithelial layer** (protects the eye against excessive incident light)

- **two muscles**
  - sphincter pupilae m. - circular fibres – miosis
  - dilatator pupilae m. – radial fibres – mydriasis
IRIS

- Supplies with major and minor vascular (arterial) circle (not visible)

- Colour of the iris – depends on the amount of pigment melanin (developing – to 6th month of life)

- At the periphery, the iris is attached to the ciliary body.
Ciliary body

- **Is continuation of the iris** - 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**

**FUNCTION:**

1. **ciliary muscle ( = accommodative muscle)** is responsible for accommodation.
2. **production of the aqueous humor**
   On the surface of ciliary body – approx. 60-80 ciliary processes (pars plicata part of ciliary body)
   Nonpigmented layer of the epithelium covering the ciliary processes produces the aqueous humor
3. **Between ciliary processes - lens zonules** (suspenory ligaments) are attached
   Suspensory ligaments are attached to the ciliary processes and the equator of the lens.
ZONULAR FIBRES  (Zonula ciliaris Zinii )

• = ”Suspensory ligaments ”

• Series of very fine transparent fibres which run from ciliary processes

• Attached to the lens capsule around equator

• Holds the lens in the right position

• Assist action of ciliary muscle during accommodation

• It is composed of glycoproteins and mucopolysaccharides
**Accommodation** = the ability of the eye to change its focus from distant to near objects (and vice versa). This process is achieved by the lens changing its shape.

**Changes during accommodation:**

A) contraction of ciliary muscles;
B) approximation of ciliary muscles to lens;
C) relaxation of suspensory ligament;
D) increased curvature of anterior surface of lens
Choroid – middle layer of the eyeball

- Contains vessels and lot of pigmented cells
- It is a dark, highly vascular layer situated in between the sclera and retina.
- Function: as “lymphatic nodule,” supplies nourishment to the outer layers of the retina
- The outer layers of retina are dependent for their nutrition (feeds pigmented epithelium of the retina and a layer of rods and cones)
- The inflammation of choroid always involves the retina.
- The blood flow through the choroid is the highest in the entire body
- 3 layers: suprachoroid / blood vessel layer / Bruch`s membrane
INFLAMMATION OF THE UVEAL TRACT (UVEITIS)
Uveitis – definition

• The term *uveitis* strictly means inflammation of the uveal tissue only.

• However, there is always associated inflammation of the adjacent structures such as retina, vitreous, sclera or cornea.
Uveitis- classification

- **Anterior** (iritis, iridocyclitis)
  - the inflammation of the iris (iritis) and pars plicata of the ciliary body (cyclitis) or iridocyclitis.
- **Intermediate** (pars planitis)
  - the inflammation of the pars plana part of the ciliary body.
- **Posterior**
  - the inflammation of the choroid (choroiditis or combination with retina as chorioretinitis - focalis, diseminnata, nekrotisans)
- **Panuveitis** - all uveal parts

- **Endophtalmitis** - inflammation of the internal structures of the eye, i.e. choroid, retina and vitreous.

- **Panophtalmitis** - all intraocular structures: purulent inflammation of all the structures of the eye. There is inflammation of all the three coats of the eye and Tenon’s capsule as well.
Types of inflammation

- **Granulomatous** - infective in nature, chronic inflammation, limited granuloma formation (less virulent, usually endogenous)

- **Non-granulomatous** - it is usually due to allergic or immune related reaction.
  - acute onset and shorter duration
  - acute diffuse damage - response to virulent pathogens, hyperergic states with a strong inflammatory and exudative response
Etiology

• In most cases, uveitis is not due to direct infection. It is usually due to allergy or hypersensitivity reaction to an infectious agent.

• **Hypersensitivity reaction**—It occurs due to hypersensitivity reaction to autologous tissue components (autoimmune reaction). Therefore uveitis occurs commonly in association with: rheumatoid arthritis, systemic lupus erythematosus, sarcoidosis, ankylosing spondylitis, Reiter’s disease, Behcet’s syndrome.
Classification - CAUSES

I. Endogenous
Organisms lodged in some other organ of the body reach the eye through the bloodstream.

- Infections - viral, bacterial, parasitic, fungal disease
- Systemic inflammatory disease (reumatoid disease)
- Idiopathic nonspecific and specific (allergic, autoimmune background)

II. Exogenous - after trauma or after surgery
It occurs due to a perforating wound or corneal ulcer. It causes acute purulent iridocyclitis and sometimes panophthalmitis.
General mechanism of ocular inflammation

The following mechanisms are involved in inflammation of the uveal tissue

- **Vessel dilatation** → deep ciliary (mixed) injection
- **Vascular permeability** → aqueous **flare** (flare in anterior chamber = cell movement), **tyndalisation**
- **Migration of cells** → keratic precipitates (cell precipitates on the corneal endothelium)
  → exudate in anterior chamber
  → hypopyon (pus level on AC bottom)
Anterior uveitis  (iridocyclitis)

**SYMPTOMS:**
- Eye-dull pain- worse at acomodation
- sensitivity to touch, increased tearing, photophobia
  (Photophobia is due to pain induced by pupillary constriction and ciliary spasm because of inflammation!)

  ( without any mucopurulent discharge )

- red eye = ciliary injection = hyperaemia around the limbus due to the dilation of anterior ciliary vessels
- decreased vision - due to exudate in AC ( anterior chamber )
Anterior uveitis (iritocyclitis)

**SIGNS:**

- Iris - decoloration or nodule formation in the iris tissue
- Inflammatory exudate - Ly, Le, fibrin
  (The exudate tends to stick to the damaged endothelium in the lower part of cornea in a triangular pattern due to the convection currents in anterior chamber and effect of gravity.)
- Tyndalisation - flare in anterior chamber - cell movement
- Cell precipitates on the corneal endothelium
- Hypopyon - pus level on AC bottom, serofibrinous
- Posterior synechiae - adhesions between the iris and lens, pupil is lobed
Anterior uveitis (iritidocyclitis)

• **Diagnosis:**
  - subjective compliants
  - biomicroscopy (at the slit lamp) - objective findings
  - laboratory exams - ASLO, LATEX, CRP, Brucela, Leptospira, BWR, HSV, HIV ...
  - **foci** - ORL, urology, gynecology, lung (tuberculin test), dentist
  - immunological examination
Therapy of the anterior uveitis (iritocyclitis)

**mydriatics** (barrier formation, break adhesions between iris and anterior capsule of the lens, reduce pain, swelling) ..... ATROPINE drops

**Corticosteroids** – anti-inflammatory, anti-allergic, anti-fibrotic activity

**anti-inflammatory drugs** - drops and pills (Ibuprofen)

**specific anti-inflammatory therapy** (antibiotics, antivirals, ...) immunomodulation, immunosuppressants

**Non-steroidal anti-inflammatory (NSAIDs)**: drops, pills, etc.

Indomethacin, diclofenac. These are safer as prolonged use of steroids which may produce complications: steroid glaucoma by reducing outflow facility, cataract and secondary infection with bacteria or fungi.
Corticosteroids

- These are anti-inflammatory in action.
- They are very useful in controlling inflammation in the acute phase.
- Due to their anti-allergic and anti-fibrotic activity they reduce fibrosis and thus prevent disorganisation and destruction of tissues.
- **Topical**—eyedrops and eye ointment
- **Subconjunctival injection**
- **Periocular injection of depot steroids**
- **Systemic steroids**
- **Intravitreal injections**
Intermediate uveitis
(CHRONIC POSTERIOR CYCLITIS OR PARS PLANITIS)

It affects the pars plana of the ciliary body and often the peripheral retina and underlying choroid.

- Idiopathic, chronic, relapsing inflammatory disease
- or associated with systemic diseases
  - multiple sclerosis (MS), sarcoidosis, Lyme disease, reumatoid arthritis
- It is common in children and young adults
- **VITREUS** is the major site of the inflammation
Intermediate uveitis
(CHRONIC POSTERIOR CYCLITIS OR PARS PLANITIS)

SYMPTOMS: Subjectiv:
- flying flies (seeing floaters)
- deterioration of central visual acuity
- blurred vision

SIGNS: Objectiv:
- cell infiltration, condensation in vitreus, vitritis
- inflammatory cells in the anterior chamber (+ or -)
- "snowballs, snowbanking, i.e. grey-white plaques involving the inferior pars plana
- cotton ball 'bearing whitish-based pars planitis
Intermediary uveitis – THERAPY

- **corticosteroids** - parabulbar (periocular) injection
  - therapeutic concentration behind the lens
  - prolonged effect
- **systemic corticosteroids**
  - *oral prednisone* (start with large dose: Prednison 1-2 mg/kg/day – maintained until clinical effect is seen, reduce slow)
  - or
  - *i.v. methylprednisolone* 1g /day 2-3 days
- **immunosuppressants**
- **intravitreal steroids**
- **vitreoretinal surgery in severe cases**
Posterior uveitis
- choroiditis or chorioretinitis

Symptoms:
• Painless - no sensitive innervation
• Calm eye - often unrecognized
• Metamorphopsia—straight line appears wavy due to oedema of the retina.
• Decrease in visual acuity- due to retinal lesions and opacities in the vitreous (floaters) and especially when macular involvement
• Sensation of flying flies, cobwebs, blurred vision, vitreous exudate (Black spots are seen floating in the vitreous (vitritis).
Posterior uveitis-chorioretinitis

**SIGNS:**
- In early stage one or more yellowish areas (yellowish gray exudative changes) with ill-defined edges are seen deep under the retinal vessels. This appearance is due to infiltration of the choroid and presence of exudates which hide the choroidal vessels.
- There may be sheathing of retinal vessels.
- Complications: macular edema, optic disc edema
- Usually chronic disease
- Healing stage - scars - fibrous tissue (whitish scar-scleral shines), pigment clumps

(yellow lesions become white due to fibrosis and the lesions are surrounded by black pigments)
Posterior uveitis

**Diagnosis:**
- Subjective problems, symptoms
- Objective findings (panfundoscopy)
- Visual field test
- Fluoroangiography, OCT (optical coherence tomography)
- Sampling of vitreous material
- Immunological exams
Posterior uveitis- therapy

- General treatment - according to etiology, antibiotics, antivirals..
- Specific treatment is required for causative organism such as: toxoplamosis, toxocarasis, tuberculosis, syphilis, etc.
- Immunosuppression
- Nonspecific anti-inflammatory therapy, Wobenzym
- Corticosteroids: systemic, parabulbar, intravitreal steroids
- Vitreoretinal surgery (pars plana vitrectomy), application of antibiotics into vitreous
Clinical types of uveitis
Sarcoidosis

- chronic multisystem granulomatous disease of unknown etiology

- predominantly affects lungs and intrathoracic lymph nodes, but any organ can be involved

- involvement of the eyes and adnexa occurs in 25-80%

- **Ocular manifestation of sarcoidosis**
  - Adnexal involvement: Orbital lacrimal gland granuloma
    - Extraocular muscles granuloma
    - Lid granuloma
    - Conjunctiva granuloma
    - Conjunctivitis
  - Episcleritis, scleritis
  - Keratitis
  - Uveitis granulomatous or non-granulomatous
    - Anterior uveitis
    - Intermediate uveitis
    - Posterior uveitis
    - Panuveitis
  - Optic nerve involvement
    - Papilitis
    - Optic disc granuloma
    - Papilledema
Toxoplasmosis = protozoan infection

- common cause of posterior uveitis
- Toxoplasma gondii

- spread by haematogenous dissemination – to the muscles, brain, choroid and retina
- clinical picture – necrotizing chorioretinitis – often self-limited, progressively resolves, leaving scar(s)

- recurrences – unpredictable

- in immunosuppressive patient – multifocal, progressive chorioretinitis
Toxocariasis

- Toxocara canis, cati
- Usually – in children
- The soil of parks and playgrounds is commonly contaminated with the eggs
- Larvae migrate through the intestine wall to the blood, encystes in various tissues (eye, brain, liver,..)
- Ocular finding – granuloma
- Death of the larva leads to severe intraocular inflammation
Fungal uveitis

- uncommon
- causative organism – Candida, Aspergillus, cryptococcus, fusarium
- progressive intraocular inflammation
- more often in: immunocompromised patients or in organ transplant patient

Th: Amphotericin-B
- Surgical: pars plana vitrectomy
**Sympathetic ophthalmia**

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

- Tissues in the injured eye (uveal tract, lens, and retina) act as antigens and provoke an autoimmune disorder in the unaffected eye.
Etiology of sympathetic ophthalmitis

• **After trauma, penetrating injury** (sympathetic eye) –
  - immune response to antigens of injured uvea: iris, ciliary body or choroid
  - autoimmune response to proteins of damaged tissue
    ---- antibodies attack uvea of the other (second) eye

• on the second (sympathizing) eye – fibrovascular chronic inflammation of the iris and ciliary body

• **Iridocyclitis or chorioretinitis**
Sympathetic ophthalmia

( prevalence 0.06-0.01%)

Clinical signs include:

- combined injections
- cells and protein in the anterior chamber and vitreous body,
- papillary and retinal edema, granulomatous inflammation of the choroid.
- The disorder has a chronic clinical course and may involve severe complications such as secondary glaucoma, secondary cataract, retinal detachment, and shrinkage of the eyeball.
- Sympathetic ophthalmia can lead to blindness in particularly severe cases
Sympathetic ophthalmia

**Therapy:**

- high doses of corticosteroids i.v., p.o., topical
- local mydriatics
- Immunosuppression
- antibiotics
- surgery (enucleation sympathetic eye) - the injured eye, which is usually blind, must be in severe cases enucleated to eliminate the antigen and to save 2-nd eye

Severe prognosis ..... without treatment......blindness!
**Endophthalmitis**
(inflammation of the internal structures of the eye, i.e. choroid, retina and vitreous)

- **Exogenous:**
  - after trauma, penetrating injuries, intraocular foreign body, perforation of suppurative corneal ulcer
  - after intraocular surgery (sources: bacterial flora of the eyelids, conjunctiva and lacrimal passage)

- **Endogenous:**
  - Systemic infection may cause metastatic infection (septic emboli), e.g. AIDS, viral fever, septicemia. It may occur in immunodeficient host and uncontrolled diabetic patients.

  (DM gangrene, focuses in the body, lowered immunity – purulent microbes - inflammation of the middle and inner layer of the eye)
Endophthalmitis – clinical features

• There is sudden onset with severe pain and redness in the eye in acute cases.
• Visual loss
• Lid oedema, chemosis and corneal haze
• Fibrinous exudate or hypopyon
• Vitritis (haze in the vitreous from inflammatory cells)
• Yellowish reflex seen behind the lens
• Inability to visualize the fundus