Arawind FAQs in Ophthalmology



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3rd Edition



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Uvea

3.1 UVEITIS—HISTORY AND CLINICAL FEATURES

1. Why is uvea named so?

"Uvea" is derived from the Greek word "*uva*" meaning grape. When the sclera is removed, the center of the eyeball appears like a grape and hence the name.

2. What does iris mean?

Iris is derived from a Greek word meaning rainbow/halo.

3. What is the normal iris pattern and what are the parts of iris?

Iris can be divided into ciliary and pupillary zones by a zigzag line called collarette.

At the pupillary margin, there is a pigmented frill-fringe of black pigment (due to slight extension of the posterior pigmented epithelium of the iris).

Pupillary zone is between the pigmented frill and collarette—relatively smooth and flat.

Ciliary zone consists of radial streaks that are straight when the pupil is small and wavy when the pupil is dilated.

4. Why do the iris and ciliary body often get involved together?

The presence of the major arterial circle causes the involvement of both the iris and ciliary body in pathological conditions. The blood supply to the choroid is essentially segmental, and hence the lesions are also isolated.

5. What is the importance of age in uveitis?

- i. Children:
 - Juvenile rheumatoid arthritis (JRA)
 - Toxocariasis
- ii. Young adults:
 - Behçet's disease
 - Human leukocyte antigen B27 (HLA-B27)-associated uveitis
 - Fuchs' uveitis
 - Sarcoidosis

- Herpes simplex
- Toxoplasmosis

iii. Middle age:

- Reiter's disease
- Ankylosing spondylitis (AS)
- Vogt-Koyanagi-Harada (VKH) syndrome
- White dot syndromes
- Toxoplasmosis

iv. Elderly individuals:

- VKH syndrome
- Herpes zoster ophthalmicus
- Tuberculosis (TB)
- Leprosy

6. What is the importance of gender in uveitis?

- i. Males:
 - AS
 - Reiter's disease
 - Behçet's disease
 - Sympathetic ophthalmia (SO)
- ii. Females:
 - Rheumatoid arthritis (RA)
 - JRA

7. What is the importance of race in uveitis?

- i. Caucasians: AS
- ii. Blacks: Sarcoidosis
- iii. Orientals: VKH syndrome
- iv. Orientals: Behçet's disease
- v. Filipinos: Coccidioidomycosis

8. Definitions.

i. Anterior uveitis:

It is subdivided into:

- *Iritis:* Inflammation involving iris only
- *Iridocyclitis:* Inflammation involving iris and anterior part of ciliary body (the pars plana)
- ii. *Intermediate uveitis*: Predominant involvement of pars plana and extreme periphery of retina
- iii. *Posterior uveitis:* Inflammation beyond the posterior border of vitreous base
- iv. Panuveitis: Involvement of the entire uveal tract
- v. *Retinochoroiditis:* Primary involvement of retina with associated involvement of choroid

- vi. *Chorioretinitis:* Primary involvement of choroid with associated involvement of retina
- vii. *Vitritis:* Presence of cells in the vitreous secondary to inflammation of uvea, retina, optic nerve, and blood vessels
- viii. Diffuse choroiditis: Generalized inflammation of the choroid
 - ix. *Disseminated choroiditis*: Two or more scattered foci of inflammation in the choroid, retina, or both
 - x. *Exogenous infection:* Infection occurring as a result of external injury to uvea, operative trauma, or any other event leading to invasion of microorganisms from outside
 - xi. *Endogenous infection:* Infection occurring as a result of microorganisms or their products released from a different site within the body
- xii. Secondary infection: Infection of uveal tract due to spread from other ocular tissue

9. How do you classify uveitis?

International Uveitis Study Group Classification

i. Anatomical classification

Term	Primary site of inflammation	Includes
Anterior uveitis	Anterior chamber (AC)	 Iritis Iridocyclitis Anterior cyclitis
Intermediate uveitis	Vitreous	 Pars planitis Posterior cyclitis Hyalitis
Posterior uveitis	Retina/choroid	ChoroiditisChorioretinitisRetinochoroiditisRetinitisNeuroretinitis
Panuveitis	AC/vitreous/retina/choroid	

ii. Clinical classification

- Infectious:
 - Bacterial
 - Viral
 - Fungal
 - Parasitic
- Noninfectious:
 - Known systemic association
 - No known systemic association

- Masquerade:
 - Neoplastic
 - Nonneoplastic

Anatomical Classification

Tessler's Classification

- i. Sclerouveitis
- ii. Keratouveitis
- iii. Anterior uveitis
- iv. Iritis
- v. Iridocyclitis
- vi. Intermediate uveitis
- vii. Cyclitis, vitritis
- viii. Pars planitis
 - ix. Posterior uveitis
 - x. Retinitis
 - xi. Choroiditis

Pathological Classification

- i. Granulomatous and nongranulomatous
- ii. Suppurative and exudative

Etiological Classification

Infectious

- i. *Exogenous: Staphylococcus, Pseudomonas, Propionibacterium acnes.* Secondary—iridocyclitis associated with herpetic keratitis, iridocyclitis associated with anterior and posterior scleritis
- ii. Endogenous:

Bacterial	 TB Syphilis Gonorrhea
Viral	Herpes simplexCytomegalovirus (CMV)MeaslesInfluenza
Fungal	HistoplasmosisCoccidioidomycosisCandidiasis
Parasitic	ToxoplasmosisToxocariasisOnchocerciasisPneumocystis carinii

Hypersensitivity/autoimmune

- i. Lens-induced—autoimmune reaction to lens protein
- ii. SO-autoimmunity to uveal pigment
- iii. VKH-suspected autoimmune origin
- iv. Behçet's

Toxic

- i. Systemic toxins—onchocercal uveitis
- ii. Endo-ocular uveitis—atrophic uveitis in degenerating eyes
- iii. Iridocyclitis in retinal detachment (RD) due to unusual proteins reaching through retinal tear
- iv. Chemical irritants—miotics and cytotoxic agents

Associated with Systemic Conditions

- i. Associated with arthritis
- ii. AS
- iii. RA
- iv. JRA
- v. Psoriatic arthritis
- vi. Associated with gastrointestinal tract (GIT) disorders: Ulcerative colitis
- vii. Associated with anergy: Sarcoidosis, leprosy, TB

Associated with Neoplasms

i. Retinoblastoma, choroidal melanoma

Idiopathic

- i. Specific-Fuchs'
- ii. Nonspecific-account for 25% of all uveitis

Occurrence of Uveitis

i. Most common type of uveitis:

Anterior uveitis is the most common type, followed by intermediate, posterior, and panuveitis

- ii. Most common age group affected, i.e., 20-40 years
- iii. Common causes of uveitis in young adults:
 - Behcet's
 - Sarcoidosis
 - Fuchs' heterochromic iridocyclitis
 - Herpes simplex
 - Toxoplasmosis

- iv. Causes of uveitis in middle ages:
 - Reiter's disease
 - AS
 - VKH syndrome
 - White dot syndrome
 - Toxoplasmosis
- v. Uveitic entities with sex predilection:

Males	Females	
AS	RA	
Reiter's syndrome	JRA	
Behçet's		·

- vi. Racial influence on uveitis
 - Caucasians: AS, Reiter's syndrome
 - Black: Sarcoidosis
 - Orientals: VKH, Behçet's
 - Filipino: Coccidioidomycosis
- vii. Geographic influence on uveitis
 - "Histoplasmosis belt" of Ohio, Missouri, and Mississippi—histoplasmosis
 - Japan and Mediterranean countries—Behçet's disease and VKH
 - San Joaquin Valley of California—coccidioidomycosis
- viii. Genetic/familial influence on uveitis
 - RA and collagen disease
 - Syphilis
 - Human immunodeficiency virus (HIV) and CMV
 - TB
 - Pars planitis

Standardization of Uveitis Nomenclature (SUN) Working Group "Activity of Uveitis" terminology:

Inactive: Grade 0 cells in anterior chamber (AC)

Worsening activity: Two-step increase in level of inflammation Improving activity: Two-step decrease in level of inflammation

Remission: Inactive disease for >3 months after discontinuing all treatment for eye disease

SUN Working Group "Descriptors in Uveitis":

Onset: Sudden/insidious

Duration:

Limited: <3 months' duration *Persistent:* >3 months' duration

Courses:

Acute: Sudden onset and limited duration

Recurrent: Repeated episodes separated by periods of inactivity without treatment for a duration of 3 months

Chronic: Persistent uveitis with relapse in <3 months after discontinuing treatment

Remission: Inactive disease for at least 3 months after discontinuing treatment

10. Name causes of acute and chronic posterior uveitis.

- i. Acute posterior uveitis occurs in toxoplasmosis
- ii. Chronic posterior uveitis occurs in pars planitis and toxocariasis

11. Name causes of acute generalized uveitis.

- i. Endophthalmitis
- ii. SO

12. What are the causes of acute suppurative uveitis?

- i. Panophthalmitis
- ii. Endophthalmitis
- iii. Suppurative iridocyclitis

13. What are the causes of unilateral nongranulomatous uveitis?

- i. Fuchs'
- ii. AS

14. What are the causes of unilateral granulomatous uveitis?

- i. Viral
- ii. Lens-induced

15. What are the causes of bilateral granulomatous uveitis?

Infectious		Noninfectious
ТВ		Sarcoidosis
Leprosy	,	Vogt–Koyanagi–Harada (VKH) syndrome
Syphilis	:	Sympathetic ophthalmia (SO)

16. What is Fuchs' heterochromic uveitis? What are the gonioscopic findings in Fuchs'? What are its sequelae?

Fuchs' uveitis is a unilateral idiopathic nongranulomatous anterior uveitis occurring in young adults. It is associated with heterochromia of the iris.

Gonioscopic finding in Fuchs'—fine filamentous vessels bridging angle Sequelae in Fuchs'—cataract

-glaucoma

17. What are the causes of uveitis associated with vitritis?

- i. Pars planitis
- ii. Irvine-Gass syndrome

- iii. Active retinitis
- iv. Trauma

18. What is the relevance of eliciting the following history in uveitis?

- i. Trauma/eye surgery—SO
- ii. Vitiligo, alopecia, poliosis-VKH syndrome
- iii. Rashes: Hyper-/hypopigmentation—leprosy
- iv. Low back pain/joint pain—AS, RA, psoriatic arthritis
- v. Painful mouth ulcers—Behçet's syndrome
- vi. Dysentery, altered bowel habits—ulcerative colitis
- vii. Ringing in the ears, hearing loss, headache—VKH
- viii. Respiratory symptoms

19. Why history of fever is important in uveitis?

- i. TB
- ii. Syphilis
- iii. Leprosy
- iv. Leptospirosis
- v. Collagen vascular disorders

20. What are the systemic findings associated with uveitis?

Skin:

- i. Rash of secondary syphilis
- ii. Erythema nodosum, sarcoidosis, Behçet's disease
- iii. Psoriasis-plaques, arthritis
- iv. Keratoderma blennorrhagica, Reiter's syndrome
- v. Kaposi sarcoma
- vi. Leprosy
- vii. VKH syndrome
- viii. Sarcoidosis

Hair:

i. Alopecia: VKH, secondary syphilis

ii. Poliosis: VKH

Nails:

- i. Pitting
- ii. Psoriasis

Dysentery:

i. Reiter's syndrome

Mouth ulcers:

i. Painful: Behçet's syndrome

ii. Painless: Reiter's syndrome

Aravind FAQs in Ophthalmology

This book is a compilation of frequently asked questions and the answers expected in postgraduate ophthalmology examination. The examination time is a tough period to any student facing it, and a quick and concise revision is required for the students to pass it. While this book is not a replacement to the standard textbooks in ophthalmology, it will boost the self-confidence of the postgraduates to face their examinations with confidence. These questions have been painstakingly gathered over a 15-year period from the collective experience of several senior teachers at Aravind Eye Hospital, Madurai, Tamil Nadu, India and the answers have been constantly refined. Apart from the exam-oriented questions, this book also contains examples of case sheet writing and different management scenarios, which will help the students to logically analyze and answer in a coherent manner.

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international journals such as Ophthalmology, American Journal of Ophthalmology, and British Journal of Ophthalmology. In addition, he serves in the Advisory Board of JAMA Ophthalmology, being the first Indian to get this honor.

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