What is uveitis?
Uveitis: Overview

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Why did the first answer include the hedge ‘in theory at least’?
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Why did the first answer include the hedge ‘in theory at least’?
Because in clinical parlance, some of the inflammatory conditions to which the term *uveitis* is applied involve non-uveal tissues (eg, the retina; vitreous).
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What three structures comprise the uvea?
The choroid, ciliary body and iris.

In a nutshell, how should the process of evaluating and managing a case of uveitis proceed?
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(We will address all of these steps through the course of this topic.)
Uveitis: Profiling

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Of these, which three are far-and-away the most important?
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**Process note:** Throughout the remainder of this slide-set, questions will drop regarding various common uveitides. Don’t despair or become frustrated if you can’t answer most of them! *The purpose of these questions is not to pimp you; rather, it is to introduce the common uveitides, and their traits.*
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**What etiologies should come to mind when encountering uveitis in:**

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What do these abbreviations stand for?
Rb:
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*Rb*: Retinoblastoma
*JXG*: Juvenile xanthogranuloma
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  - Masquerade syndrome: (4) Intraocular lymphoma; uveal melanoma; OIS; mets
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**What do these abbreviations stand for?**
- **TINU**: Tubulointerstitial nephritis and uveitis
- **JRA**: Juvenile rheumatoid arthritis (aka juvenile idiopathic arthritis)
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**What specifically constitute the ‘germane aspects’ of a pt’s identity?**
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--Gender
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**How specific does the age need to be?**
Age can be considered in terms of stages of life; ie, infancy, childhood, adolescence, young adulthood, elderly

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What does FHI stand for?

Fuchs heterochromic iridocyclitis

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What does OIS stand for?

Ocular ischemic syndrome
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--Age
--Gender
--Ethnicity/race
--Geographic history
--Family history
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What etiology should come first to mind when encountering uveitis in person of the following ethnicities:
--Native American?
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What specifically constitute the ‘germane aspects’ of a pt’s identity?

--Age
--Gender
--Ethnicity/race
--Geographic history
--Family history
--Vocational/avocational activities

What etiology should come first to mind when encountering uveitis in person of the following ethnicities:

--**Native American**? Vogt-Koyanagi-Harada dz (VKH)

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- Age
- Gender
- Ethnicity/race
- Geographic history
- Family history
- Social history

**What etiology should come first to mind when encountering uveitis in person of the following ethnicities:**
- **Native American?** Vogt-Koyanagi-Harada dz (VKH)
- **African-American?**

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**Uveitis: Profiling**

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--Age
--Gender
--Ethnicity/race
--Geographic history
--Family history
--Sociocultural activities

**What etiology should come first to mind when encountering uveitis in person of the following ethnicities:**

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--**African-American**? Sarcoid
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What all is encompassed by geographic history?

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--Gender
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What all is encompassed by geographic history?
The pt’s country of origin, country of residence, and recent travel history

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What etiology should come first to mind when encountering uveitis in person from:

--Central/South America?

--The Eastern Mediterranean?

--Turkey?

--The Middle East?

--China?

--Japan?

--India?

--Africa?

--The Northeast US?

--Wisconsin?

--The Ohio River /Mississippi River valleys?

--Hawaii?

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--Central/South America? Cysticercosis
--The Eastern Mediterranean? Behçet
--Turkey? Behçet
--The Middle East? Behçet
--China? (2)

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Uveitis: **Profiling**

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Behçet disease is most common amongst people living along the ancient ‘Silk Road’ stretching from the Eastern Mediterranean to the Eastern Pacific rim.
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--The Ohio River /Mississippi River valleys? Ocular histoplasmosis syndrome (POHS)

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---Family history
- Social history

What are you looking for in the family history?
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What are you looking for in the family history?
In addition to any history of uveitis, it’s important to inquire about infectious contacts, as well as autoimmune diseases.
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- Age
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**What are you looking for in the social history?**

- Sexual activities that place the pt at risk for infection
- Dietary habits that do the same (eg, consuming raw meat)
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**What etiology should come first to mind when encountering uveitis in person who:**
--- Eats raw/undercooked meat?
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--Dietary habits that do the same (eg, consuming raw meat)  
--Recreational drug use, especially intravenous

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**What etiology should come first to mind when encountering uveitis in person who:**

--**Eats raw/undercooked meat?** Toxoplasmosis

---

**What is the goal of the profiling process?**

A concise declarative statement that captures the important features of the case; eg, ‘Ms. Jones is a 40 y.o. female of Middle-Eastern descent who presents with a chronic bilateral granulomatous panuveitis associated with headache and tinnitus.’
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**Social history**
--Sexual activities that place the pt at risk for infection
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--Recreational drug use, especially intravenous

What etiology should come first to mind when encountering uveitis in person who:
--Eats raw/undercooked meat? Toxoplasmosis
--Has an IVDU history?
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What etiology should come first to mind when encountering uveitis in person who:

--- **Eats raw/undercooked meat?** Toxoplasmosis
--- **Has an IVDU history?** Candida

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**Vocational/avocational activities**

**What are you looking for regarding vocational/avocational activities?**
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--Vocational/avocational activities

What are you looking for regarding vocational/avocational activities?
Exposure to farm animals; mining work; sewage exposure (eg, plumbers); pet ownership
Uveitis: Profiling

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1) Germaine aspects of the pt’s personal history, demographics, etc;

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3) certain key features of the inflammation itself (eg, location, duration, etc).

What is the goal of the profiling process?
A concise declarative statement that captures the important features of the case; eg, ‘Ms. Jones is a 40 y.o. female of Middle-Eastern descent who presents with a chronic bilateral granulomatous panuveitis associated with headache and tinnitus.’

What specifically constitute the ‘germaine aspects’ of a pt’s identity?

- Age
- Gender
- Ethnicity/race
- Geographic history
- Family history
- Social history

What are you looking for regarding vocational/avocational activities?

Exposure to farm animals; mining work; sewage exposure (eg, plumbers); pet ownership

What etiology should come first to mind when encountering uveitis in person who:

-- Works with cattle?
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**Exposure to farm animals**; mining work; sewage exposure (eg, plumbers); pet ownership

What etiology should come first to mind when encountering uveitis in person who:
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**What etiology should come first to mind when encountering uveitis in person who:**
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- Age
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What needs to be covered in the ROS/PE?

Everything!!!! --Constitutional--Neuro--ENT:--Skin--Pulmonary--Cardiac--GI--GU--Musculoskeletal
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That said, there are a number of particular ‘positives’ you need to be on the lookout for…

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--Constitutional
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What constitutional S/S should you be looking for?

--Fever:
  (3) Reiter syndrome; infectious endophthalmitis; polyarteritis nodosa (PAN)
--Night sweats:
  (2) TB; malignancy
--Flulike prodrome:
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--Constitutional
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What neuro S/S should you be looking for?
--Headache
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--Meningismus
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--Cranial neuropathies

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--Headache:
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What neuro S/S should you be looking for? What etiology should come first to mind when encountering uveitis in a person with a positive finding for each?
--Headache: VKH
--Tinnitus: VKH
--Meningismus: VKH
--Hearing loss: VKH
--Psychiatric/cognitive manifestations: VKH; Behçet; multiple sclerosis (MS)
--Cranial neuropathies: MS; Lyme; Whipple’s; PAN
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Via a careful Review of Systems and Physical Exam.

What is the goal of the profiling process?
A concise declarative statement that captures the important features of the case; eg, ‘Ms. Jones is a 40 y.o. female of Middle-Eastern descent who presents with a chronic bilateral granulomatous panuveitis associated with headache and tinnitus.’

What needs to be covered in the ROS/PE? Everything!!!!
--Constitutional
--Neuro

What neuro S/S should you be looking for? What etiology should come first to mind when encountering uveitis in a person with a positive finding for each?

--Headache: VKH
--Tinnitus: VKH
--Meningismus:
--Hearing loss
--Psychiatric/cognitive manifestations
--Cranial neuropathies

--VKH; Behçet; multiple sclerosis (MS)
--(2) MS; Lyme

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--- **Whipple’s**

--- **Polyarteritis nodosa (PAN)**
**Uveitis: Profiling**

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--Constitutional
--Neuro
--ENT
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--External ear inflammation
--Saddle-nose deformity
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--External ear inflammation:
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What **ENT S/S** should you be looking for? What etiology should come first to mind when encountering uveitis in a person with a positive finding for each?

---**External ear inflammation**: Relapsing polychondritis (RP)
---**Saddle-nose deformity**: (3) RP; Wegener’s; congenital syphilis
---**Sinusitis**: Wegener’s
---**Oral ulcers**:
    ----Alone:
    ----Along with genital ulcers: (2)
In the context of uveitis, to what does ‘profiling’ refer? It refers to identifying three components of a uveitis case: 1) Germaine aspects of the pt’s personal history, demographics, etc; 2) nonocular signs and symptoms associated with the uveitis; and 3) certain key features of the inflammation itself (eg, location, duration, etc).

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--Oral ulcers:
----Alone: Inflammatory bowel dz (IBD)
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In what fundamental way does the presentation of oral ulcers in Behçet differ from that of reactive arthritis? The oral ulcers in one are extremely painful, whereas those in the other are painless.
In the context of uveitis, to what does ‘profiling’ refer? It refers to identifying three components of a uveitis case:
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How does one go about identifying associated signs and symptoms? Via a careful review of systems and physical exam.

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--Saddle-nose deformity: (3) RP; Wegener’s; congenital syphilis

--Sinusitis: Wegener’s
--Oral ulcers:----Alone: Inflammatory bowel dz (IBD)
----Along with genital ulcers: (2) Behçet; reactive arthritis

What sign, if present, is particularly worrisome for Wegener’s sinusitis? If the associated nasal discharge is bloody.
In the context of uveitis, to what does ‘profiling’ refer? It refers to identifying three components of a uveitis case:
1) Germaine aspects of the pt’s personal history, demographics, etc;
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What is the goal of the profiling process? A concise declarative statement that captures the important features of the case; eg, ‘Ms. Jones is a 40 y.o. female of Middle-Eastern descent who presents with a chronic bilateral granulomatous panuveitis associated with headache and tinnitus.’

How does one go about identifying associated signs and symptoms? Via a careful Review of Systems and Physical Exam

What needs to be covered in the ROS/PE? Everything!!!!
--Constitutional
--Neuro
--ENT
--Skin
--Pulmonary
--Cardiac
--GI
--GU

What ENT S/S should you be looking for? What etiology should come first to mind when encountering uveitis in a person with a positive finding for each?
--External ear inflammation: Relapsing polychondritis (RP)
--Saddle-nose deformity: (3) RP; Wegener’s; congenital syphilis
--Sinusitis: Wegener’s

What sign, if present, is particularly worrisome for Wegener’s sinusitis? If the associated nasal discharge is bloody, are particularly worrisome for Wegener’s sinusitis.

Uveitis: Profiling

1) The uveitis is profiled
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That said, there are a number of particular ‘positives’ you need to be on the lookout for…

What skin S/S should you be looking for?
--Rash:
----‘Bull’s eye’: Lyme
----Palms/soles: (3) Syphilis; Reactive arthritis; psoriatic arthritis
--Poliosis: VKH
--Madarosis: VKH
--Vitiligo: VKH
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What is the palm/sole rash of reactive arthritis called?
Keratoderma blennorrhagicum
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**--Erythema nodosum:** Behçet

How does erythema nodosum typically present?

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--Erythema nodosum: Behçet

How does erythema nodosum typically present?
As tender purple nodules on the pre-tibial skin.
In the context of uveitis, to what does ‘profiling’ refer? It refers to identifying three components of a uveitis case: 1) germane aspects of the pt’s personal history, demographics, etc; 2) nonocular signs and symptoms associated with the uveitis; and 3) certain key features of the inflammation itself (eg, location, duration, etc).

What is the goal of the profiling process? A concise declarative statement that captures the important features of the case; eg, ‘Ms. Jones is a 40 y.o. female of Middle-Eastern descent who presents with a chronic bilateral granulomatous panuveitis associated with headache and tinnitus.’

How does one go about identifying associated signs and symptoms? Via a careful **Review of Systems** and **Physical Exam**.

What needs to be covered in the ROS/PE? **Everything!!!!**

--- Constitutional
--- Neuro
--- ENT
--- Skin
--- Pulmonary
--- Cardiac
--- GI
--- GU
--- Musculoskeletal

What pulmonary S/S should you be looking for?

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--Constitutional
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What pulmonary S/S should you be looking for?

--SOB/DOE
--Cough
--Hemoptysis

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1) The uveitis is profiled
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What pulmonary S/S should you be looking for? What etiology should come first to mind when encountering uveitis in a person with a positive finding for each?

--SOB/DOE: (2)
--Cough
--Hemoptysis

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**Uveitis: Profiling**

In the context of uveitis, to what does ‘profiling’ refer?

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**What pulmonary S/S should you be looking for? What etiology should come first to mind when encountering uveitis in a person with a positive finding for each?**

- **SOB/DOE**: (2) Sarcoid; TB
- Cough
- Hemoptysis

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--SOB/DOE: (2) Sarcoid; TB
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--Hemoptysis: (2) Wegener’s; TB

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**Uveitis: Profiling**

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Via a careful **Review of Systems** and **Physical Exam**

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**What cardiac S/S should you be looking for?**

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What cardiac S/S should you be looking for?

--‘Cardiac involvement’

The BCSC Uveitis book doesn’t go into much detail concerning specific cardiac signs/symptoms (with two exceptions to be noted shortly)

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--‘Cardiac involvement’: (5)
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--‘Cardiac involvement’: (5) Behçet; sarcoid; Lyme; SLE; PAN

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With regard to specific cardiac-related complaints, the BCSC book mentions pericarditis as a manifestation of three of these conditions. Which ones?
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Behçet, SLE and PAN
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The BCSC book also mentions heart block/conduction issues as a manifestation of one of these conditions. Which one?
Uveitis: Profiling

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What GI S/S should you be looking for? --Diarrhea
--Ulcers

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What GI S/S should you be looking for? What etiology should come first to mind when encountering uveitis in a person with a positive finding for each?

--Diarrhea: (2)
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--Diarrhea: (2) IBD; Whipple’s
--Ulcers

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-- Constitutional
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What GU S/S should you be looking for?
--
--
--

That said, there are a number of particular points you need to be on the lookout for:

What GU S/S should you be looking for?
-- Genital lesions:
  -- Alone: Syphilis
  -- Along with oral ulcers: Behçet; Reiter’s
-- Urethral discharge:
  -- Syphilis; Reiter’s
-- Nephritis:
  -- Wegener’s; TINU

Uveitis:
Profiling

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What GU S/S should you be looking for? What etiology should come first to mind when encountering uveitis in a person with a positive finding for each?

--Genital lesions:
----Alone:
----Urethral discharge
----Nephritis
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--Genital lesions:
----Alone: Syphilis

--Urethral discharge
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--Genital lesions:
----Behçet; reactive arthritis
----Alone:

--Urethral discharge
--Syphilis; Reiter’s

--Nephritis
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What is the penile lesion of reactive arthritis called? Circinate balanitis.
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---Nephritis: (2) Wegener’s; TINU

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-- Arthralgias:
   ---- Low back pain: HLA-B27 dz, especially ankylosing spondylitis
   ---- Distal phalangeal pain/swelling: HLA-B27 dz, especially psoriatic arthritis
   ---- The knee: Lyme; JIA; reactive arthritis; Behçet
   ---- Ankle: Reactive arthritis
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Two words
Uveitis: Profiling

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The goal is to identify the etiology and the differential diagnosis.

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What is the tasty term for the swollen digits encountered in psoriatic arthritis?
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Sausage digits
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1) **Germane aspects of the pt’s personal history, demographics, etc;**
2) **nonocular signs and symptoms associated with the uveitis;** and
3) **certain key features of the inflammation itself (eg, location, duration, etc)**

What is the goal of the profiling process?

A concise declarative statement that captures the important features of the case; eg, ‘Ms. Jones is a 40 y.o. female of Middle-Eastern descent who presents with a chronic bilateral granulomatous panuveitis associated with headache and tinnitus.’

How does one go about identifying associated signs and symptoms? Via a careful **Review of Systems and Physical Exam**

What needs to be covered in the ROS/PE? **Everything!!!!**

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It refers to identifying (ie, profiling) two components of a uveitis case:
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2) Certain key features of the inflammation itself (eg, location, duration, etc)

‘What precisely constitutes ‘certain key features of the inflammation itself’? is the subject of the next section…

A concise declarative statement that captures the important features of the case; eg, ‘Ms. Jones is a 40 y.o. female of Middle-Eastern descent who presents with a chronic bilateral granulomatous panuveitis.’
Uveitis: Classification

*In the context of uveitis, what does the acronym SUN stand for?*

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The onset, duration and course of the uveitis
--Onset: something vs something
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--Course: something vs something vs ...

How are onset, duration and course delineated?

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--Abb. & two words
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...is divided into three subtypes based on what attribute?

**Cell location**

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--Anterior?

--Intermediate

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**Anterior uveitis**

*If cell is located…*

- Exclusively in the AC
  - It is called: Iritis

- In the AC and the anterior vitreous
  - It is called: ???

- Exclusively in the AVit
  - It is called: Anterior cyclitis

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When inflammatory cells aggregate in small clumps in the far periphery, what are these aggregates called?

- Snowballs
- When the aggregates cover a much larger area of the far periphery, what are they called?
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Intermediate uveitis
…Is divided into two subtypes based on what attribute?
Whether it is idiopathic, vs associated with an infection or systemic inflammatory condition

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Intermediate uveitis

If condition is…

Idiopathic

Not idiopathic

It is called:

?
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*Pars planitis*

Intermediate uveitis

*If condition is…*

**Idiopathic**

**Not idiopathic**

*It is called:*

- Pars planitis

*With respect to uveitis: Which study is performed next?*

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Intermediate uveitis
If condition is...

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Posterior uveitis

…is divided into three subtypes based on what attribute?

Inflammation location

It is called:  It is called:  It is called:

?  ?  ?

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What are the three ‘inflammation locations’?

Posterior uveitis

If inflammation is located…

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- **Severity** of the uveitis:
  - AC cell grade
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Posterior uveitis

If inflammation is located…

- Exclusively in the choroid
- In both the choroid and the retina
- Exclusively in the retina

What are the three ‘inflammation locations’?

- It is called: ?
- It is called: ?
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If inflammation is located…

Exclusively in the choroid
It is called:

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Posterior uveitis

If inflammation is located…

Exclusively in the choroid

It is called: Choroiditis

In both the choroid and the retina

It is called: ?

Exclusively in the retina

It is called: ?

1) The uveitis is profiled
2) The profiled case is meshed
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In the context of uveitis, what does the acronym SUN stand for?

**SUN** stands for Standardization of Uveitis Nomenclature, a working group appointed by the International Ocular Inflammation Society.

What was the task of the SUN Working Group?

The task of the SUN Working Group was to standardize uveitis nomenclature.

The SUN classification system is based on three sets of criteria:

**1. Location of the uveitis**
   - Anterior
   - Intermediate
   - **Posterior?**
   - Panuveitis

**2. Onset, duration, and course of the uveitis**
   - Onset: Sudden vs insidious
   - Duration: Limited vs persistent
   - Course: Acute vs recurrent vs chronic

**3. Severity of the uveitis**
   - AC cell grade
   - AC flare grade
   - Vitreous haze score

Posterior uveitis

If inflammation is located…

- Exclusively in the choroid
  - **It is called:** Choroiditis

- In both the choroid and the retina
  - **It is called:** ?

- Exclusively in the retina
  - **It is called:** ?

With respect to uveitis: Where is the primary location of inflammation in…

- **Anterior uveitis**? The anterior chamber (although cell ‘spillover’ into the anterior vitreous may occur)
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The severity of the uveitis
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--Vitreous haze score

Posterior uveitis

If inflammation is located…

Exclusively in the choroid

It is called:

Choroiditis

In both the choroid and the retina

It is called:

?

Exclusively in the retina

It is called:

Retinitis

With respect to uveitis: Where is the primary location of inflammation in…

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Posterior uveitis

If inflammation is located…

Exclusively in the choroid

It is called:

Choroiditis

In both the choroid and the retina

It is called:

? (Transition state or not clearly defined)

Exclusively in the retina

It is called:

Retinitis

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Posterior uveitis

If inflammation is located...

- Exclusively in the choroid
  - It is called: Choroiditis
- In both the choroid and the retina
  - It is called: Chorioretinitis or Retinochoroiditis
- Exclusively in the retina
  - It is called: Retinitis

With respect to uveitis: Where is the primary location of inflammation in...

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  The main vitreous cavity, +/- the peripheral retina and/or pars plana

- **Posterior uveitis**?
  The retina, choroid, and/or optic nerve head

**Posterior uveitis**

*If inflammation is located…*

- Exclusively in the choroid
  - It is called: **Choroiditis**

- In both the choroid and the retina
  - It is called: **Neuroretinitis**

- Exclusively in the retina
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**Panuveitis?**
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- **Panuveitis**? All three locations are equally involved

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**Panuveitis**

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- Onset: Sudden vs insidious
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- Course: Acute vs recurrent vs chronic

**The severity of the uveitis**
- AC cell grade
- AC flare grade
- Vitreous haze score

A pt has dense AC cell, scant anterior vitreous cell, and cystoid macular edema. Given all three locations are involved, this pt has panuveitis, yes?

With respect to uveitis, where is the primary inflammation located in…

**Anterior uveitis?** The anterior chamber (although cell ‘spillover’ into the anterior vitreous may occur).

**Intermediate uveitis?** The vitreous, peripheral retina and/or pars plana

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A pt has dense AC cell, scant anterior vitreous cell, and cystoid macular edema. Given all three locations are involved, this pt has panuveitis, yes?
No. The description clearly suggests the pt has an anterior uveitis with spillover of cell into the anterior vitreous, along with cystoid macular edema caused by the anterior uveitis. In other words, despite the fact that all three locations are involved, the primary location is anterior, making this an anterior uveitis.

With respect to uveitis, where is the primary inflammation located in…

Anterior uveitis? The anterior chamber (although cell ‘spillover’ into the anterior vitreous may occur)
Intermediate uveitis? The vitreous, peripheral retina and/or pars plana
Posterior uveitis? The retina and/or choroid
Panuveitis? All three locations are equally involved

Uveitis: Classification

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With respect to uveitis, what is the difference between... Sudden and insidious onset?
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With respect to uveitis, what is the difference between…
Sudden and insidious onset? Sudden-onset uveitis presents with abrupt development of symptoms (pain, photophobia) and signs (injection); whereas insidious uveitis is largely sign- and symptom-free
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Sudden and insidious onset? Sudden-onset uveitis presents with abrupt development of symptoms (pain, photophobia) and signs (injection); whereas insidious uveitis is largely sign- and symptom-free

Limited and persistent duration? Limited lasts < 3 months; persistent > 3 months
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Limited and persistent duration? Limited lasts < 3 months; persistent > 3 months
Acute, recurrent and chronic course? Acute uveitis comes on suddenly and resolves fairly quickly. Recurrent uveitis eventually relapses, but is quiescent off-treatment for at least 3 months. Chronic uveitis also relapses, but its quiescent periods off-treatment last less than 3 months.
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How is the severity of the AC cellular reaction graded?
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How is the severity of the AC cellular reaction graded? By counting the number of WBCs visible within a 1 x 1 mm slit-lamp beam

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Where/what is Arlt’s triangle?
It’s a triangle with its apex at the corneal center and base near the inferior border of the cornea

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  - Heterochromia

Broadly, what sorts of iris changes must be looked for?

In a pt with uveitis and heterochromia, which iris is more likely to be the abnormal one--the lighter iris, or the darker?
In the context of uveitis, what does the acronym SUN stand for?
Standardization of Uveitis Nomenclature, a working group appointed by the International Ocular Inflammation Society

What was the task of the SUN Working Group?
To standardize uveitis nomenclature

The SUN classification system is based on three sets of criteria. What are they?

<table>
<thead>
<tr>
<th>The <strong>location</strong> of the uveitis</th>
<th>The <strong>onset, duration</strong> and <strong>course</strong> of the uveitis</th>
<th>The <strong>severity</strong> of the uveitis</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Anterior</td>
<td>- Onset: Sudden vs insidious</td>
<td>- AC cell grade</td>
</tr>
<tr>
<td>- Intermediate</td>
<td>- Duration: Limited vs persistent</td>
<td>- AC flare grade</td>
</tr>
<tr>
<td>- Posterior</td>
<td>- Course: Acute vs recurrent vs chronic</td>
<td>- Vitreous haze score</td>
</tr>
<tr>
<td>- Panuveitis</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

In addition to location, onset/duration/course, and severity, what other aspects of presentation are important in classifying a case of uveitis?

- Laterality (ie, whether it is uni- vs bilateral)
- The character of keratic precipitates (KP) if present
- Iris changes
- Heterochromia

Broadly, what sorts of iris changes must be looked for?

- Atrophy
- Synechiae
- Nodules
- Heterochromia

In a pt with uveitis and heterochromia, which iris is more likely to be the abnormal one--the lighter iris, or the darker? The lighter