Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral, or bilateral disease?
Is SO a unilateral, or bilateral disease?

It is a **bilateral** condition.
Q

- Is SO a unilateral, or bilateral disease?
  *It is a **bilateral** condition*

- SO is a bilateral **general condition** following ocular injury or different event in one eye
Is SO a unilateral, or bilateral disease?
It is a **bilateral** condition
SO is a bilateral **panuveitis** following ocular **injury** or **surgery** in one eye
Is SO a unilateral, or bilateral disease? It is a **bilateral** condition.

SO is a bilateral **panuveitis** following ocular injury or surgery.

Is it granulomatous, or nongranulomatous?
Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral, or bilateral disease? **It is a bilateral condition.**
- SO is a bilateral panuveitis following ocular injury or surgery.

*Is it granulomatous, or nongranulomatous?*

**Granulomatous**
Is SO a unilateral, or bilateral disease?
It is a **bilateral** condition

SO is a bilateral **panuveitis** following ocular **injury or surgery** in one eye

*Which is a more common cause: Injury, or surgery?*
Is SO a unilateral, or bilateral disease?
It is a **bilateral** condition

SO is a bilateral **panuveitis** following ocular **injury** or **surgery** in one eye

Which is a more common cause: Injury, or surgery?
Back in the day, it was injury by a mile. However, refinements in managing ocular trauma have reduced the post-trauma SO rate such that it is now below that after surgery
**Sympathetic Ophthalmia (SO): Wadda ya know?**

- Is SO a unilateral, or bilateral disease? It is a **bilateral** condition.
- SO is a bilateral panuveitis following ocular **injury or surgery** in one eye.

*Which is a more common cause: Injury, or surgery?*
Back in the day, it was injury by a mile. However, refinements in managing ocular trauma have reduced the post-trauma SO rate such that it is now below that after surgery.

*What is the classic injury-related scenario associated with SO; ie, what would be seen at the slit-lamp?*
Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral, or bilateral disease?
  It is a **bilateral** condition

- SO is a bilateral **panuveitis** following ocular **injury or surgery** in one eye

Which is a more common cause: Injury, or surgery?
Back in the day, it was injury by a mile. However, refinements in managing ocular trauma have reduced the post-trauma SO rate such that it is now below that after surgery

What is the classic injury-related scenario associated with SO; ie, what would be seen at the slit-lamp?
- tissue incarcerated in the wound
Sympathetic Ophthalmia (SO): Wadda ya know?

- **Is SO a unilateral, or bilateral disease?**
  - It is a **bilateral** condition
- **SO is a bilateral panuveitis following ocular injury or surgery** in one eye

Which is a more common cause: Injury, or surgery?
Back in the day, it was injury by a mile. However, refinements in managing ocular trauma have reduced the post-trauma SO rate such that it is now below that after surgery

What is the classic injury-related scenario associated with SO; ie, what would be seen at the slit-lamp?
- Uveal tissue incarcerated in the wound
Q

Is SO a unilateral, or bilateral disease?
It is a bilateral condition

SO is a bilateral panuveitis following ocular injury or surgery in one eye

You know that perforating/penetrating injury can result in SO, but what about a contusion?
Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral, or bilateral disease?
  It is a **bilateral** condition
- SO is a bilateral panuveitis following ocular injury or surgery in one eye

You know that perforating/penetrating injury can result in SO, but what about a contusion? Yes, globe contusion has resulted in SO (although this is **exceedingly** rare)
Is SO a unilateral, or bilateral disease?
It is a bilateral condition.

SO is a bilateral panuveitis following ocular injury or surgery.

Which of these surgeries has been associated with SO--
--PPV?
--Cataract surgery?
--Trabeculectomy?
--Cyclocryoablation?
--Cyclophotocoagulation?
**A**

- **Is SO a unilateral, or bilateral disease?** It is a **bilateral** condition.

- **SO is a bilateral panuveitis following ocular injury or surgery.**

**Sympathetic Ophthalmia (SO): Wadda ya know?**

Which of these surgeries has been associated with SO--

-- PPV!
-- Cataract surgery!
-- Trabeculectomy!
-- Cyclocryoablation!
-- Cyclophotocoagulation!

All have been associated with SO!
Q

- Is SO a unilateral, or bilateral disease? 
  It is a **bilateral** condition.
- SO is a bilateral panuveitis following ocular injury or surgery.

**Sympathetic Ophthalmia (SO): Wadda ya know?**

Which surgery has been associated with SO--
- PPV?
- Cataract surgery?
- Trabeculectomy?
- Cyclocryoablation?
- Cyclophotocoagulation?

Which surgery carries the highest risk of SO?
Is SO a unilateral, or bilateral disease? It is a **bilateral** condition.

SO is a bilateral panuveitis following ocular injury or surgery.

Which of these surgeries has been associated with SO—\textbf{PPV}!

- Cataract surgery
- Trabeculectomy
- Cyclocryoablation
- Cyclophotocoagulation

Which surgery carries the highest risk of SO? \textbf{Vitrectomy}
Q

- Is SO a unilateral, or bilateral disease? It is a **bilateral** condition.
- SO is a bilateral panuveitis following ocular injury or surgery.

**Sympathetic Ophthalmia (SO): Wadda ya know?**

Which of these surgeries has been associated with SO--

---PPV!

Which incidence is higher: Post-PPV endophthalmitis, or post-PPV SO?

---Cyclophotocoagulation
Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral, or bilateral disease?
  - It is a **bilateral** condition.
- SO is a bilateral panuveitis following ocular injury or surgery.

Which of these surgeries has been associated with SO--

- PPV!

Which incidence is higher: Post-PPV endophthalmitis, or post-PPV SO?

- Post-PPV SO

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Is SO a unilateral, or bilateral disease? **It is a bilateral condition**

SO is a bilateral **panuveitis** following ocular injury or surgery in one eye

- Previously injured/operated eye is called the **inciting eye**
Is SO a unilateral, or bilateral disease?

It is a **bilateral** condition

SO is a bilateral **panuveitis** following ocular **injury** or **surgery** in one eye

- Previously injured/operated eye is called the **exciting** eye
Is SO a unilateral, or bilateral disease?

It is a **bilateral** condition.

SO is a bilateral **panuveitis** following ocular injury or surgery in one eye.

- Previously injured/operated eye is called the **exciting** eye.
- Previously normal eye is called the **sympathizing** eye.
Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral, or bilateral disease?
  It is a **bilateral** condition

- SO is a bilateral **panuveitis** following ocular injury or surgery in one eye
  - Previously injured/operated eye is called the **exciting** eye
  - Previously normal eye is called the **sympathizing** eye
● Is SO a unilateral, or bilateral disease? It is a **bilateral** condition

● SO is a bilateral **panuveitis** following ocular injury or surgery in one eye
  ● Previously injured/operated eye is called the **exciting** eye
  ● Previously normal eye is called the **sympathizing** eye

● What is the cause of SO?
Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral, or bilateral disease? It is a **bilateral** condition.
- SO is a bilateral **panuveitis** following ocular **injury** or **surgery** in one eye.
  - Previously injured/operated eye is called the **exciting** eye.
  - Previously normal eye is called the **sympathizing** eye.
- What is the cause of SO? **Unknown**.
Is SO a unilateral, or bilateral disease? It is bilateral.

SO is a bilateral panuveitis following ocular injury or surgery in one eye. Previously injured/operated eye is called the exciting eye. Previously normal eye is called the sympathizing eye.

What is the cause of SO? Unknown

That said, we are not completely in the dark vis a vis etiology, so…

Is it infectious, or noninfectious? Most experts are convinced it is noninfectious.

Ok, so it’s immune-related. Is it T-cell, or B-cell mediated? This one we know—T-cells are the culprit.

To what antigens are the T cells responding? This is less certain, but it seems uveal antigens—specifically, melanin-associated?—play a central role. 
Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral or bilateral disease?
  - It is bilateral.
  - SO is a bilateral panuveitis following ocular injury or surgery in one eye.
  - Previously injured/operated eye is called the exciting eye.
  - Previously normal eye is called the sympathizing eye.

- What is the cause of SO? Unknown

That said, we are not completely in the dark vis a vis etiology, so…

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To what antigens are the T cells responding?

This is less certain, but it seems uveal antigens—specifically, melanin-associated?—play a central role

What is the cause of SO? Unknown
Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral, or bilateral disease?
  - It is bilateral

- SO is a bilateral panuveitis following ocular injury or surgery in one eye
  - Previously injured/operated eye is called the exciting eye
  - Previously normal eye is called the sympathizing eye

- What is the cause of SO? Unknown

That said, we are not completely in the dark vis a vis etiology, so…

**Is it infectious, or noninfectious?**
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Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral, or bilateral disease? It is a bilateral condition.
- SO is a bilateral panuveitis following ocular injury or surgery in one eye. Previously injured/operated eye is called the exciting eye. Previously normal eye is called the sympathizing eye.
- What is the cause of SO? Unknown

That said, we are not completely in the dark vis a vis etiology, so…

_Is it infectious, or noninfectious?_ Most experts are convinced it is noninfectious

_Ok, so it’s immune-related. Is it T-cell, or B-cell mediated?_ This one we know--T-cells are the culprit

_To what antigens are the T cells responding?_
Is SO a unilateral, or bilateral disease? It is a bilateral condition.

SO is a bilateral panuveitis following ocular injury or surgery in one eye. Previously injured/operated eye is called the exciting eye; previously normal eye is called the sympathizing eye.

What is the cause of SO? Unknown

That said, we are not completely in the dark vis a vis etiology, so...

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To what antigens are the T cells responding? This is less certain, but it seems uveal antigens--specifically, melanin-associated?--play a central role.
Q

Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral, or bilateral disease? 
  It is a **bilateral** condition
- SO is a bilateral **panuveitis** following ocular injury or surgery in one eye
  - Previously injured/operated eye is called the **exciting** eye
  - Previously normal eye is called the **sympathizing** eye
- What is the cause of SO? **Unknown**
- What is the classic presenting complaint?
Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral, or bilateral disease?
  - It is a **bilateral** condition

- SO is a bilateral **panuveitis** following ocular **injury** or **surgery** in one eye
  - Previously injured/operated eye is called the **exciting** eye
  - Previously normal eye is called the **sympathizing** eye

- What is the cause of SO? **Unknown**

- What is the classic presenting complaint?
  - Impaired near vision (ie, loss of accommodation) in the previously normal eye
Is SO a unilateral, or bilateral disease? It is a **bilateral** condition.

SO is a bilateral panuveitis following ocular **injury** or **surgery** in one eye.
- Previously injured/operated eye is called the **exciting** eye.
- Previously normal eye is called the **sympathizing** eye.

What is the cause of SO? Unknown.

What is **the classic presenting complaint**?
Impaired near vision (ie, **loss of accommodation**).

*Take note of this!* IMHO, it would make a great Orals question--a pt with a remote hx of ocular trauma presenting with what sounds like a simple case of early-onset presbyopia. Would be all too easy to send him/her out with OTC readers, thereby consigning this hypothetical pt to profound vision loss--and yourself to repeating the Boards.
SO: Treatment/course/prognosis
- In a nutshell, treat like any other
SO: Treatment/course/prognosis

- In a nutshell, treat like any other panuveitis
Sympathetic Ophthalmia (SO): Wadda ya know?

- SO: Treatment/course/prognosis
  - In a nutshell, treat like any other panuveitis
    - Local
Sympathetic Ophthalmia (SO): Wadda ya know?

- SO: Treatment/course/prognosis
  - In a nutshell, treat like any other panuveitis
    - Local steroids
Sympathetic Ophthalmia (SO): Wadda ya know?

SO: Treatment/course/prognosis

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - For comfort
SO: Treatment/course/prognosis

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
Sympathetic Ophthalmia (SO): Wadda ya know?

SO: Treatment/course/prognosis

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic
SO: Treatment/course/prognosis

- In a nutshell, treat like any other **panuveitis**
  - Local **steroids**
  - **Cycloplegia** for comfort
  - +/- systemic **steroids**

Sympathetic Ophthalmia (SO): Wadda ya know?
SO: Treatment/course/prognosis

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic steroids
  - May need systemic immunomodulation/suppression
SO: Treatment/course/prognosis

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic steroids
  - May need systemic immunomodulation/suppression

Sympathetic Ophthalmia (SO): Wadda ya know?
Sympathetic Ophthalmia (SO): Wadda ya know?

SO: Treatment/course/prognosis

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic steroids
  - May need systemic immunomodulation/suppression
  - Should the exciting eye be removed?
SO: Treatment/course/prognosis

- In a nutshell, treat like any other **panuveitis**
  - Local **steroids**
  - **Cycloplegia** for comfort
  - +/- systemic **steroids**
  - May need systemic **immunomodulation/suppression**
  - Should the exciting eye be removed? **Not if it has any potential for useful vision**
Sympathetic Ophthalmia (SO): Wadda ya know?

**SO: Treatment/course/prognosis**

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic steroids
  - May need systemic immunomodulation/suppression
  - Should the exciting eye be removed? **Not if it has any potential for useful vision**

*Can the risk of SO be reduced by prophylactic enucleation?*
SO: Treatment/course/prognosis

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic steroids
  - May need systemic immunomodulation/suppression
- Should the exciting eye be removed? Not if it has any potential for useful vision

Can the risk of SO be reduced by prophylactic enucleation?
Yes, if it’s done within about [time period] of injury occurrence
SO: Treatment/course/prognosis

- In a nutshell, treat like any other *panuveitis*
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic steroids
  - May need systemic *immunomodulation/suppression*
  - **Should the exciting eye be removed?** Not if it has any potential for useful vision

*Can the risk of SO be reduced by prophylactic enucleation?*
Yes, if it’s done within about 2 weeks of injury occurrence
**Sympathetic Ophthalmia (SO): Wadda ya know?**

**SO: Treatment/course/prognosis**
- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic steroids
  - May need systemic immunomodulation/suppression
- Should the exciting eye be removed? Not if it has any potential for useful vision

*Can the risk of SO be reduced by prophylactic enucleation?*
Yes, if it’s done within about 2 weeks of injury occurrence

*Once the process commences, can the course (severity; duration, etc) be mitigated by enucleating the exciting eye?*
SO: Sympathetic Ophthalmia (SO): Wadda ya know?

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic steroids
  - May need systemic immunomodulation/suppression
- Should the exciting eye be removed? Not if it has any potential for useful vision

Can the risk of SO be reduced by prophylactic enucleation?
Yes, if it’s done within about 2 weeks of injury occurrence

Once the process commences, can the course (severity, duration, etc) be mitigated by enucleating the exciting eye?
No
SO: Treatment/course/prognosis

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic steroids
  - May need systemic immunomodulation/suppression
  - Should the exciting eye be removed? Not if it has any potential for useful vision

Q

Sympathetic Ophthalmia (SO): Wadda ya know?

- Can the risk of SO be reduced by prophylactic enucleation?
  Yes, if it’s done within about 2 weeks of injury occurrence

- If the injured eye is not salvageable, is evisceration a reasonable option?
  No

- Once the process commences, can the course (severity, duration, etc) be mitigated by enucleating the exciting eye?
  No
SO: Treatment/course/prognosis

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic steroids
  - May need systemic immunomodulation/suppression
  - Should the exciting eye be removed? Not if it has any potential for useful vision

---

Sympathetic Ophthalmia (SO): Wadda ya know?

**Can the risk of SO be reduced by prophylactic enucleation?**
- Yes, if it’s done within about 2 weeks of injury occurrence.

**Once the process commences, can the course (severity, duration, etc) be mitigated by enucleating the exciting eye?**
- No

**If the injured eye is not salvageable, is evisc a reasonable option?**
- Tough call, but probably not. Evisc incurs a risk of leaving minute amounts of uveal tissue, and thus does not completely eliminate the risk of inciting SO.
SO: Treatment/course/prognosis

- In a nutshell, treat like any other **panuveitis**
  - Local **steroids**
  - **Cycloplegia** for comfort
  - +/- systemic **steroids**
  - May need systemic **immunomodulation/suppression**
  - Should the exciting eye be removed? **Not if it has any potential for useful vision**

- What is the course? **chronic vs a one-time acute event**

**Sympathetic Ophthalmia (SO): Wadda ya know?**
SO: Treatment/course/prognosis

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic steroids
  - May need systemic immunomodulation/suppression
  - Should the exciting eye be removed? Not if it has any potential for useful vision
- What is the course? Chronic--waxing and waning

Sympathetic Ophthalmia (SO): Wadda ya know?
SO: Treatment/course/prognosis

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic steroids
  - May need systemic immunomodulation/suppression
  - Should the exciting eye be removed? Not if it has any potential for useful vision

- What is the course? Chronic--waxing and waning
- What is the prognosis?
SO: Treatment/course/prognosis

- In a nutshell, treat like any other panuveitis
  - Local steroids
  - Cycloplegia for comfort
  - +/- systemic steroids
  - May need systemic immunomodulation/suppression
  - Should the exciting eye be removed? Not if it has any potential for useful vision

- What is the course? Chronic--waxing and waning

- What is the prognosis? Far better than it used to be—with modern steroids/immuno drugs, probability of maintaining useful vision is excellent
Fill in the blanks regarding the time course of post-trauma SO:

- The onset of SO is very rare before [length of time since trauma].
Fill in the blanks regarding the time course of post-trauma SO:

- The onset of SO is very rare before ____2 weeks____.
Sympathetic Ophthalmia (SO): Wadda ya know?

- **Fill in the blanks regarding the time course of post-trauma SO:**

  - The onset of SO is very rare before __________.
  - 80% declare by __________, and 90% by __________.
Fill in the blanks regarding the time course of post-trauma SO:

- The onset of SO is very rare before **2 weeks**.
- 80% declare by **3 months**, and 90% by **1 year**.
**Fill in the blanks regarding the time course of post-trauma SO:**

- The onset of SO is very rare before **2 weeks**.
- 80% declare by **3 months**, and 90% by **1 year**.
- The shortest documented time to onset was **[length of time since trauma]**; the longest, **[length of time since trauma]**.
Fill in the blanks regarding the time course of post-trauma SO:

- The onset of SO is very rare before **2 weeks**.
- 80% declare by **3 months**, and 90% by **1 year**.
- The shortest documented time to onset was **5 days**; the longest, **66 years**.
Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

- Is VKH a unilateral, or bilateral disease?
Is VKH a unilateral, or bilateral disease?

It is a **bilateral** condition.
Is VKH a unilateral, or bilateral disease?

It is a **bilateral** condition

VKH is a bilateral **absent** ocular or **history**
Is VKH a unilateral, or bilateral disease?

It is a bilateral condition

VKH is a bilateral panuveitis absent ocular injury or surgery history
Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

- Is VKH a unilateral, or bilateral disease? It is a **bilateral** condition.
- VKH is a bilateral **panuveitis** absent ocular injury or surgery history.

**Is it granulomatous, or nongranulomatous?**
Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

- Is VKH a unilateral, or bilateral disease? It is a **bilateral** condition.
- VKH is a bilateral **panuveitis** absent ocular injury or surgery history.

**Is it granulomatous, or nongranulomatous?**

Granulomatous
Is VKH a unilateral, or bilateral disease?

It is a **bilateral** condition.

VKH is a bilateral **panuveitis** **absent** ocular **injury** or **surgery** history.

What is the cause of VKH?
Is VKH a unilateral, or bilateral disease?
It is a **bilateral** condition

VKH is a bilateral **panuveitis** **absent** ocular **injury** or **surgery** history

What is the cause of VKH? **Unknown**
Is VKH a unilateral, or bilateral disease?
It is a **bilateral** condition

- VKH is a bilateral panuveitis **absent** ocular injury or **surgery** history

- **What is the cause of VKH?** *Unknown*

That said, we are not completely in the dark vis a vis etiology, so...

*Is it infectious, or noninfectious?*
- Is VKH a unilateral, or bilateral disease? It is a **bilateral** condition.
- VKH is a bilateral **panuveitis** **absent** ocular injury or surgery history.
- What is the cause of VKH? **Unknown**

That said, we are not completely in the dark vis a vis etiology, so…

*Is it infectious, or noninfectious?*
Most experts are convinced it is noninfectious.
Is VKH a unilateral, or bilateral disease?
It is a \textbf{bilateral} condition

VKH is a bilateral \textit{panuveitis} \textit{absent} ocular injury or \textit{surgery} history

What is \textbf{the cause of VKH}? \textbf{Unknown}

That said, we are not completely in the dark vis a vis etiology, so…

\textit{Is it infectious, or noninfectious?}
Most experts are convinced it is noninfectious

\textit{Ok, so it’s immune-related. Is it T-cell, or B-cell mediated?}
Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

- Is VKH a unilateral, or bilateral disease? It is a bilateral condition.
- VKH is a bilateral panuveitis absent ocular injury or surgery history.
- What is the cause of VKH? Unknown.

That said, we are not completely in the dark vis a vis etiology, so…

*Is it infectious, or noninfectious?* 
Most experts are convinced it is noninfectious.

*Ok, so it’s immune-related. Is it T-cell, or B-cell mediated?*  
This one we know--T-cells are the culprit.
Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

- Is VKH a unilateral, or bilateral disease?
  It is a bilateral condition
- VKH is a bilateral panuveitis absent ocular injury or surgery history
- What is the cause of VKH? Unknown

That said, we are not completely in the dark vis a vis etiology, so...

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Most experts are convinced it is noninfectious

*Ok, so it’s immune-related. Is it T-cell, or B-cell mediated?*
This one we know--T-cells are the culprit

*To what antigens are the T cells responding?*
Q/A Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

- Is VKH a unilateral, or bilateral disease? It is a bilateral condition.
- VKH is a bilateral panuveitis absent ocular injury or surgery history.
- What is the cause of VKH? Unknown.

That said, we are not completely in the dark vis a vis etiology, so...

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Most experts are convinced it is noninfectious.

*Ok, so it’s immune-related. Is it T-cell, or B-cell mediated?*
This one we know--T-cells are the culprit.

*To what antigens are the T cells responding?*
The experimental evidence strongly suggests that the antigens in question are associated with type of cell.
Is VKH a unilateral, or bilateral disease? It is a bilateral condition.

VKH is a bilateral panuveitis absent ocular injury or surgery history.

What is the cause of VKH? Unknown.

That said, we are not completely in the dark vis a vis etiology, so…

*Is it infectious, or noninfectious?*
Most experts are convinced it is noninfectious.

*Ok, so it’s immune-related. Is it T-cell, or B-cell mediated?*
This one we know--T-cells are the culprit.

*To what antigens are the T cells responding?*
The experimental evidence strongly suggests that the antigens in question are associated with melanocytes.
Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

- Is VKH a unilateral, or bilateral disease? It is a bilateral condition.
- VKH is a granulomatous panuveitis.
- What is the cause of VKH? Unknown (but T-cells, melanin implicated).

Note the similarities…

Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral, or bilateral disease? It is a bilateral condition.
- SO is a granulomatous panuveitis.
- What is the cause of VKH? Unknown (but T-cells, melanin implicated).
Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

- Is VKH a unilateral, or bilateral disease? It is a **bilateral** condition.
- VKH is a granulomatous panuveitis **absent** ocular injury/surgery history.
- What is the cause of VKH? Unknown (but T-cells, melanin implicated).

Note the similarities...And the **key difference**

Sympathetic Ophthalmia (SO): Wadda ya know?

- Is SO a unilateral, or bilateral disease? It is a **bilateral** condition.
- SO is a granulomatous panuveitis **following** ocular injury/surgery in one eye.
- What is the cause of VKH? Unknown (but T-cells, melanin implicated).
Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

- VKH: Treatment/course/prognosis
  - In a nutshell, treat like any other...
Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

- VKH: Treatment/course/prognosis
  - In a nutshell, treat like any other panuveitis
Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

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- What is the course?
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What is the course? Chronic—waxing and waning

What is the prognosis? Far better than it used to be—with modern steroids/immuno drugs, **probability of maintaining useful vision is good.**

What factors tend to limit the prognosis in VKH?
The sequelae of the severe intraocular inflammation that characterizes the disease

What are some of these sight-threatening sequelae?
- Secondary angle-closure glaucoma
- Cataracts that are difficult to manage successfully due to intra- and/or post-op complications
- Retinal neovascularization
- Phthisis
Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

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  - What is the course? **Chronic--waxing and waning**
  - What is the prognosis? **Far better than it used to be—with modern steroids/immuno drugs, probability of maintaining useful vision is good.**

Re treatment, etc: Again, note the similarities!

Sympathetic Ophthalmia (SO): Wadda ya know?

- **SO: Treatment/course/prognosis**
  - In a nutshell, treat like any other panuveitis
    - Local steroids
    - Cycloplegia for comfort
    - +/- systemic steroids
    - May need systemic immunomodulation/suppression
    - Should the exciting eye be removed? **Not if it has any potential for useful vision**
  - What is the course? **Chronic--waxing and waning**
  - What is the prognosis? **Far better than it used to be—with modern steroids/immuno drugs, probability of maintaining useful vision is excellent**
**VKH tends to present stepwise, in three stages. What are they?**

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*(If you’re thinking ‘But VKH has four stages!’…We’ll get to that momentarily.)*
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*The above bears repeating! Generally speaking:*
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*The above bears repeating!* Generally speaking: VKH starts off by inflaming the CNS.
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*The above bears repeating!* Generally speaking: VKH starts off by inflaming the CNS; then it inflames the eyes.
### Stage vs. SO vs VKH

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*The above bears repeating!* Generally speaking: **VKH starts off by inflaming the CNS**; then it **inflames the eyes**; and finally **goes about depigmenting everything**
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*How much time typically passes between the onset of the Prodromal phase and the onset of the Acute Uveitic phase?*
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How much time typically passes between the onset of the **Acute Uveitic phase and onset of the Convalescent phase**?
### Stage	| ‘Phase Phrase’	| Signs/Symptoms |
| --- | --- | --- |
| **Prodromal**  
(a couple of days) | ‘Meningo-encephalitic’ | Flu-like symptoms plus multiple and varied neurologic findings |
| **Acute Uveitic**  
(a couple of weeks) | ‘Ophthalmic’  
Depigmentation | The classic bilateral |
| **Convalescent** |  | How much time typically passes between the onset of the Acute Uveitic phase and onset of the Convalescent phase?  
A couple of weeks  
Depigmentation  
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### Stage

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Prodromal

(A couple of days)

**Acute U**

(A couple of weeks)

**Convalescent**

‘Depigmentation’

Gradual depigmentation of the skin, hair, and choroid
### Stage | ‘Phase Phrase’ | Signs/Symptoms
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**Prodromal** | ‘Meningo-encephalitic’ | Flu-like symptoms plus multiple and varied neurologic findings
(a couple of days) |  |
**Acute U** |  | *What is the most common neurologic manifestation at this stage?*
Auditory difficulties, especially tinnitus
(a couple of weeks) |  |
**Convalescent** | ‘Depigmentation’ | Gradual depigmentation of the skin, hair, and choroid
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A SO vs VKH
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What is the *classic* retinal manifestation at this stage?
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**What is the classic retinal manifestation at this stage?**
Multifocal serous retinal detachments

**Can these coalesce to form a substantial exudative retinal detachment?**
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*What is the classic retinal manifestation at this stage?*
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*Can these coalesce to form a substantial exudative retinal detachment?*
Yes
### Stage 'Phase Phrase' Signs/Symptoms

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**What is the classic retinal manifestation at this stage?**
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**Can these coalesce to form a substantial exudative retinal detachment?**
Yes

**Optic nerve edema--common, or uncommon in this phase?**
## Stage & ‘Phase Phrase’ Signs/Symptoms

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### Acute Uveitic

**What is the classic retinal manifestation at this stage?**
Multifocal serous retinal detachments

**Can these coalesce to form a substantial exudative retinal detachment?**
Yes

**Optic nerve edema--common, or uncommon in this phase?**
Very common
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**What is the classic retinal manifestation at this stage?**

Multifocal serous retinal detachments

**How does this appear on FA?**

Early, as multiple scattered pinpoint leakages; late, as pooling within the serous-RD spaces

**Optic nerve edema--common, or uncommon in this phase?**

Very common
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**Multifocal serous retinal detachments**

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### Stage vs SO vs VKH

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**What is the classic retinal manifestation at this stage?**

- Multifocal serous retinal detachments

**How does this appear on FA?**

- Early, as multiple scattered pinpoint leakages
- Late, as pooling within the serous RD spaces

**By what colorful monikers is this appearance known?**

- Starry Night Sign
- Milky Way Sign

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**Optic nerve edema**—common, or uncommon in this phase?

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### Stage vs VKH

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**Prodromal**

What is the classic retinal manifestation at this stage?

Multifocal serous retinal detachments

How does this appear on FA?

Early, as multiple scattered pinpoint leakages; late, as pooling within the serous RD spaces

By what colorful monikers is this appearance known?

It is called ‘starry night sign’ or ‘Milky Way sign’
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What is the classic retinal manifestation at this stage? Multifocal serous retinal detachments

How does this appear on FA? Early, as multiple scattered pinpoint leakages; late, as pooling within the serous RD spaces

Can SO present with multifocal serous RD and a ‘starry night’ FA? Yes

By what colorful monikers is this appearance known? It is called ‘starry night sign’ or ‘Milky Way sign’
## Stage 'Phase Phrase' Signs/Symptoms

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How does this appear on FA?

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By what colorful monikers is this appearance known?

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Can SO present with multifocal serous RD and a 'starry night' FA?

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Depigmentation of the choroid leads to a characteristic **orange-red** color change of the posterior pole. What is the classic term used in describing this finding?
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Depigmentation of the choroid leads to a characteristic **orange-red color change of the posterior pole.** What is the classic term used in describing this finding? **‘Sunset glow fundus’**
As should be apparent by now, VKH has three broad sorts of manifestations: Neurologic, Ocular and Integumentary (ie, skin and hair). These categories provide the basis for defining three “forms” of VKH.

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If a pt presents with ocular signs/symptoms **only**, s/he has…
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*If a pt presents with ocular signs/symptoms only, s/he has…Probable VKH*
As should be apparent by now, VKH has three broad sorts of manifestations: Neurologic, Ocular and Integumentary (i.e., skin and hair). These categories provide the basis for defining three “forms” of VKH.

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If a pt presents with ocular signs/symptoms only, s/he has…Probable VKH
If s/he presents with ocular plus neuro or skin/hair changes, s/he has…

(‘or’ here indicates ‘not both’)
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If a pt presents with ocular signs/symptoms only, s/he has…**Probable** VKH
If s/he presents with ocular plus neuro or skin/hair changes, s/he has…**Incomplete** VKH
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If s/he presents with all three, s/he has...
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VKH can have a fourth phase. What is it, and why does it occur?
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**VKH** can have a fourth phase. What is it, and why does it occur?
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VKH can have a fourth phase. What is it, and why does it occur?

What specific signs/symptoms characterize this stage?
**Stage** | **‘Phase Phrase’** | **Signs/Symptoms**
--- | --- | ---
*Prodromal*  
(a couple of days) | ‘Meningo-encephalitic’ | Flu-like symptoms plus multiple and varied neurologic findings

*Acute Uveitic*  
(a couple of weeks) | ‘Ophthalmic’ | The classic bilateral granulomatous panuveitis

*Convalescent* | ‘Depigmentation’ | Gradual depigmentation of the skin, hair, and choroid

*Chronic Recurrent* | | Episodes of granulomatous anterior uveitis

VKH can have a fourth phase. What is it, and why does it occur?

What specific signs/symptoms characterize this stage?
As the *Uveitis* book puts it, “The numerous clinical and pathological similarities between SO and VKH syndrome suggest that they share a similar immunopathogenesis.” Given this, it likely won’t surprise you that:

Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

Sympathetic Ophthalmia (SO): Wadda ya know?
As the *Uveitis* book puts it, “The numerous clinical and pathological similarities between SO and VKH syndrome suggest that they share a similar immunopathogenesis.” Given this, it likely won’t surprise you that:

**Number one on the DDx for VKH:** SO.

**Number one on the DDx for SO:** VKH.

Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

Sympathetic Ophthalmia (SO): Wadda ya know?
As the *Uveitis* book puts it, “The numerous clinical and pathological similarities between SO and VKH syndrome suggest that they share a similar immunopathogenesis.” Given this, it likely won’t surprise you that:

**Number one on the DDx for VKH:** **SO.**  
**Number one on the DDx for SO:** **VKH.**  
**So let’s compare and contrast them!**
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50:
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

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For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

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- More common in whites: **SO**
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- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common:
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

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How common are skin (and hair) findings in VKH?

Vitiligo

Depigmentation of the skin
For each of the following statements, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH), or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
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How common are skin (and hair) findings in VKH? They appear in about 1/3 of patients.
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH), or both.

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What is the classic skin finding?

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**SO vs VKH**

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What is the classic skin finding? Vitiligo
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What is vitiligo? Depigmentation of the skin

Vitiligo is not limited to the skin--a classic eye location is the perilimbal region.
SO vs VKH

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What is the classic skin finding? Vitiligo

What is vitiligo? Depigmentation of the skin

Vitiligo is not limited to the skin--a classic eye location is the perilimbal region. What is the eponymous name for this sign?
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH), or both:

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH

**SO vs VKH**

- How common are skin (and hair) findings in VKH?
  - They appear in about 1/3 of patients

- What is the classic skin finding?
  - Vitiligo

- What is vitiligo?
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Vitiligo is not limited to the skin--a classic eye location is the perilimbal region.

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Sugiura's sign
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH), or both.

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- More common in whites: SO
- Concurrent skin findings common: VKH

How common are skin (and hair) findings in VKH? They appear in about 1/3 of patients

What is the classic skin finding? Vitiligo

What is vitiligo? Depigmentation of the skin

Vitiligo is not limited to the skin--a classic eye location is the perilimbal region. What is the eponymous name for this sign? Sugiura’s sign

In which pt population is it common?
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20–50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**

### How common are skin (and hair) findings in VKH?
They appear in about 1/3 of patients.

### What is the classic skin finding?
Vitiligo

### What is vitiligo?
Depigmentation of the skin

Vitiligo is not limited to the skin--a classic eye location is the perilimbal region.

### What is the eponymous name for this sign?
Sugiura's sign

### In which pt population is it common?
Pts of Japanese ancestry--almost all of them will manifest this finding.
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

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Does it occur in Caucasian pts?
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- More common in whites: 
  - SO

- Concurrent skin findings common: 
  - VKH

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A ‘perilimbal adjacent’ structure also becomes depigmented. Which one?
  - The trabecular meshwork

What is the eponymous name for TM depigmentation in VKH?
  - Ohno's sign
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH), or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH

**SO vs VKH**

*How common are skin (and hair) findings in VKH?*
*They appear in about 1/3 of patients*

*What is the classic skin finding?*
*Vitiligo*

*What is vitiligo?*
*Depigmentation of the skin*

Vitiligo is not limited to the skin--a classic eye location is the perlimbal region.

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How common are skin (and hair) findings in VKH? They appear in about 1/3 of patients.

What is the classic skin finding?
Vitiligo

What is vitiligo?
Depigmentation of the skin

What are the classic hair findings?
Alopecia and poliosis

What do these terms mean?
Alopecia: Loss of hair
Poliosis: Loss of hair pigmentation
For each of the following conditions, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH

**SO vs VKH**

How common are skin (and hair) findings in VKH? They appear in about 1/3 of patients.

What is the classic skin finding? Vitiligo

What are the classic hair findings? Alopecia and poliosis
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

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How common are skin (and hair) findings in VKH? They appear in about 1/3 of patients.

What is the classic skin finding? Vitiligo

What are the classic hair findings? Alopecia and poliosis

What do these terms mean? Alopecia: Poliosis:
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH), or both.

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**How common are skin (and hair) findings in VKH?**
They appear in about 1/3 of patients.

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For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

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How common are skin (and hair) findings in VKH? They appear in about 1/3 of patients.

What is the classic skin finding? Vitiligo.

What is vitiligo? Depigmentation.

Do the skin and hair findings precede, coincide with, or follow the onset of eye inflammation in VKH?
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH), or both.

- Predilection for adults age 20–50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**

How common are skin (and hair) findings in VKH? They appear in about 1/3 of patients.

What is the classic skin finding? **Vitiligo**

What is vitiligo? **Depigmentation**

Do the skin and hair findings precede, coincide with, or follow the onset of eye inflammation in VKH? The skin/hair changes generally follow the onset of eye inflammation by a period of **weeks**.
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
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How common are skin (and hair) findings in VKH? They appear in about 1/3 of patients.

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What is vitiligo? Depigmentation

Do the skin and hair findings precede, coincide with, or follow the onset of eye inflammation in VKH? The skin/hair changes generally follow the onset of eye inflammation by a period of weeks.

Remember the pattern:
First: Neurologic
Second: Eye
Third: Integument
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- **Concurrent skin findings** commonly occur in **VKH**, **SO**?

Do skin findings occur in SO as well?
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- **Concurrent skin findings** are common: **VKH**, **SO**

Do skin findings occur in SO as well? Yes, but they are much less common.
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare:
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
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For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- **CNS involvement rare**: in **VKH**

What is the classic sign/symptoms of CNS involvement in VKH?
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- **CNS involvement rare**: in **VKH**

**Q/A**

What is the classic sign/symptoms of CNS involvement in VKH?

Auditory issues, particularly **tinnitus** and **dysacusis**.
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

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What is the classic sign/symptoms of CNS involvement in VKH?
Auditory issues, particularly tinnitus and dysacusis

*In what other ways can CNS involvement manifest?*
--
--
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

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- Concurrent skin findings common: **VKH**
- **CNS involvement rare:** in **VKH**

*What is the classic sign/symptoms of CNS involvement in VKH?*
Auditory issues, particularly tinnitus and dysacusis

*In what other ways can CNS involvement manifest?*
--Meningeal signs/symptoms (eg, nuchal rigidity)
--Headache
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
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**What is the classic sign/symptoms of CNS involvement in VKH?**
Auditory issues, particularly tinnitus and dysacusis

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**Do focal neurologic signs/symptoms occur?**
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

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- Concurrent skin findings common: VKH
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What is the classic sign/symptoms of CNS involvement in VKH?
Auditory issues, particularly tinnitus and dysacusis

In what other ways can CNS involvement manifest?
--Meningeal signs/symptoms (eg, nuchal rigidity)
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Do focal neurologic signs/symptoms occur?
They can, but are rare
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

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**What is the classic sign/symptoms of CNS involvement in VKH?**
Auditory issues, particularly tinnitus and dysacusis

**In what other ways can CNS involvement manifest?**
--Meningeal signs/symptoms (eg, nuchal rigidity)
--Headache

**Do focal neurologic signs/symptoms occur?**
They can, but are rare

**Can VKH cause an optic neuropathy?**
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- **CNS involvement rare**: in **VKH**

**What is the classic sign/symptoms of CNS involvement in VKH?**
Auditory issues, particularly tinnitus and dysacusis

**In what other ways can CNS involvement manifest?**
--Meningeal signs/symptoms (eg, nuchal rigidity)
--Headache

**Do focal neurologic signs/symptoms occur?**
They can, but are rare

**Can VKH cause an optic neuropathy?**
Yes
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**?

**Does CNS involvement occur in SO as well?**
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**? Yes!

Does CNS involvement occur in SO as well? Yes, but it is much less common
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present:
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH
- CNS involvement rare: SO
- Dalen-Fuchs nodules present: Both
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH
- CNS involvement rare: SO
- **Dalen-Fuchs nodules present:** Both

*What are Dalen-Fuchs nodules?*
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- **Dalen-Fuchs nodules present:** Both

*What are Dalen-Fuchs nodules?*
Focal collections of inflammatory cells between Bruch’s membrane and the RPE
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- **Dalen-Fuchs nodules present:** Both

**What are Dalen-Fuchs nodules?**
Focal collections of inflammatory cells between Bruch’s membrane and the RPE

*The classic test-association for Dalen-Fuchs nodules is SO/VKH. However, there is another condition, vastly more common, in which they are seen as well. So, whereas during an exam your first response to the term Dalen-Fuchs nodule should be SO/VKH, what condition should come to mind first if you encounter them in the clinic?*
For each of the following, state whether it is associated with sympathetic ophthalimia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH
- CNS involvement rare: SO
- Dalen-Fuchs nodules present: Both

What are Dalen-Fuchs nodules?
Focal collections of inflammatory cells between Bruch’s membrane and the RPE

The classic test-association for Dalen-Fuchs nodules is SO/VKH. However, there is another condition, vastly more common, in which they are seen as well. So, whereas during an exam your first response to the term Dalen-Fuchs nodule should be SO/VKH, what condition should come to mind first if you encounter them in the clinic?
Sarcoid
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- Choriocapillaris is spared:
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH
- CNS involvement rare: SO
- Dalen-Fuchs nodules present: Both
- Choriocapillaris is spared: SO
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- **Choriocapillaris is spared**: **SO**

What is the choriocapillaris?
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- **Choriocapillaris is spared: SO**

*What is the choriocapillaris?*
The innermost portion of the choroid; it is a layer of highly-fenestrated capillaries.
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH
- CNS involvement rare: SO
- Dalen-Fuchs nodules present: Both
- **Choriocapillaris is spared:** SO

A word of clarification is in order. Much is made about the presence/absence of choriocapillaris involvement as a feature distinguishing between SO and VKH. And the choriocapillaris *is* spared in SO, and it *is* affected in VKH. But note that the choriocapillaris is spared during the Acute Uveitic phase of VKH--it’s only later, during the Chronic Recurrent phase, that it becomes involved. Thus, choriocapillaris status does not distinguish between SO and VKH so much as it distinguishes between the different stages of VKH.
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- Choriocapillaris is spared: **SO**
- More common in Hispanics, Asians:
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH
- CNS involvement rare: SO
- Dalen-Fuchs nodules present: Both
- Choriocapillaris is spared: SO
- More common in Hispanics, Asians: VKH
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH
- CNS involvement rare: SO
- Dalen-Fuchs nodules present: Both
- Choriocapillaris is spared: SO
- More common in Hispanics, Asians: VKH

What other ethnic groups are frequently affected?
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH
- CNS involvement rare: SO
- Dalen-Fuchs nodules present: Both
- Choriocapillaris is spared: SO
- More common in Hispanics, Asians: VKH

What other ethnic groups are frequently affected? Native-Americans; Middle Easterners
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- Choriocapillaris is spared: **SO**
- More common in **Hispanics, Asians**: **VKH**

*What other ethnic groups are frequently affected?*
Native-Americans; Middle Easterners

*Hispanics, Asians, Native-Americans, Middle Easterners--what is the common thread?*
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH
- CNS involvement rare: SO
- Dalen-Fuchs nodules present: Both
- Choriocapillaris is spared: SO
- More common in Hispanics, Asians: VKH

What other ethnic groups are frequently affected? Native-Americans; Middle Easterners

Hispanics, Asians, Native-Americans, Middle Easterners--what is the common thread? A relatively high level of skin pigmentation
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- Choriocapillaris is spared: **SO**
- More common in **Hispanics, Asians; VKH** and Africans?

Given this, it must follow that sub-Saharan Africans are at high risk, yes?

What is the common thread?
A relatively high level of skin pigmentation
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- Choriocapillaris is spared: **SO**
- More common in Hispanics, Asians: **but not Africans**

**Given this, it must follow that sub-Saharan Africans are at high risk, yes? You’d think so, but no--their relative risk is quite low**

What is the common thread? A relatively **high level of skin pigmentation**
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
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- Dalen-Fuchs nodules present: **Both**
- Choriocapillaris is spared: **SO**
- More common in **Hispanics, Asians**; **but not Africans**

Given this, it must follow that sub-Saharan Africans are at high risk, yes? You’d think so, but no--their relative risk is quite low.

What does indicate about the disease process?

A relatively **high level of skin pigmentation**
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH
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- Dalen-Fuchs nodules present: Both
- Choriocapillaris is spared: SO
- More common in Hispanics, Asians: but not Africans

Given this, it must follow that sub-Saharan Africans are at high risk, yes? You’d think so, but no--their relative risk is quite low.

What does indicate about the disease process? It indicates that pigmentation is not the sole contributor to the disease process, but...
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH
- CNS involvement rare: SO
- Dalen-Fuchs nodules present: Both
- Choriocapillaris is spared: SO

**Speaking of contributing to the dz process…Is there a genetic predisposition to VKH?**

- Given this, it must follow that sub-Saharan Africans are at high risk, yes?
  - You’d think so, but no—their relative risk is quite low

- What does indicate about the disease process?
  - It indicates that pigmentation is not the sole contributor to the dz process

- Is the common thread?
  - A relatively high level of skin pigmentation

- Speaking of contributing to the dz process…Is there a genetic predisposition to VKH?
  - Very much so—strong HLA associations exist. All are of the HLA-DR* type—HLA-DR4 in Japanese pts; DR1 and DR4 among Hispanics; DRB among others.
  - Can these HLA types be used to differentiate between VKH and SO?
    - No, because SO has virtually identical HLA associations.
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
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**Speaking of contributing to the dz process…** Is there a genetic predisposition to VKH? Very much so--strong HLA associations exist. All are of the HLA-DR* type--HLA-DR4 in Japanese pts; DR1 and DR4 among Hispanics; DRB among others.

What does indicate about the disease process? It indicates that pigmentation is not the sole contributor to the dz process, but

**common thread?** A relatively high level of skin pigmentation
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- Choriocapillaris is spared: **SO**

**Speaking of contributing to the dz process...** *Is there a genetic predisposition to VKH?* Very much so--strong HLA associations exist. All are of the HLA-DR* type--HLA-DR4 in Japanese pts; DR1 and DR4 among Hispanics; DRB among others.

*Can these HLA types be used to differentiate between VKH and SO?*

*What does indicate about the disease process?* It indicates that pigmentation is not the sole contributor to the dz process.

*Contributor to the dz process* is the common thread?

A relatively high level of skin pigmentation
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- Choriocapillaris is spared: **SO**

**SO vs VKH**

- What other ethnic groups are frequently affected? Native-Americans; Middle Easterners

**Hispanics, Asians, Native-Americans, Middle Easterners—what is the common thread?**

A relatively high level of skin pigmentation

**Speaking of contributing to the dz process…Is there a genetic predisposition to VKH?** Very much so—strong HLA associations exist. All are of the HLA-DR* type—HLA-DR4 in Japanese pts; DR1 and DR4 among Hispanics; DRB among others.

**Can these HLA types be used to differentiate between VKH and SO?** Alas no, because SO has virtually identical HLA associations

**What does indicate about the disease process?** It indicates that pigmentation is not the sole contributor to the dz process
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- Choriocapillaris is spared: **SO**
- More common in Hispanics, Asians: **VKH**
- No gender predilection:
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- Choriocapillaris is spared: **SO**
- More common in Hispanics, Asians: **VKH**
- No gender predilection: **Depends**
  - **VKH: M = F**
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- Choriocapillaris is spared: **SO**
- More common in Hispanics, Asians: **VKH**
- No gender predilection: **Depends…**
  - **VKH:** M < F
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- Choriocapillaris is spared: **SO**
- More common in Hispanics, Asians: **VKH**
- No gender predilection: **Depends**…
  - **VKH**: M < F
  - **SO** after surgery: M = F
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH
- CNS involvement rare: SO
- Dalen-Fuchs nodules present: Both
- Choriocapillaris is spared: SO
- More common in Hispanics, Asians: VKH
- No gender predilection: Depends…
  - VKH: M < F
  - SO after surgery: M = F
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: **VKH**
- More common in whites: **SO**
- Concurrent skin findings common: **VKH**
- CNS involvement rare: **SO**
- Dalen-Fuchs nodules present: **Both**
- Choriocapillaris is spared: **SO**
- More common in Hispanics, Asians: **VKH**
- No gender predilection: **Depends**…
  - **VKH**: $M < F$
  - **SO** after surgery: $M = F$
  - **SO** after trauma: $M \neq F$
For each of the following, state whether it is associated with sympathetic ophthalmia (SO), Vogt-Koyanagi-Harada (VKH) disease, or both.

- Predilection for adults age 20 – 50: VKH
- More common in whites: SO
- Concurrent skin findings common: VKH
- CNS involvement rare: SO
- Dalen-Fuch's nodules present: Both
- Choriocapillaris is spared: SO
- More common in Hispanics, Asians: VKH
- No gender predilection: Depends…
  - VKH: M < F
  - SO after surgery: M = F
  - SO after trauma: M > F
Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?

Recall this slide from earlier in the set. As stated then, tops on the DDx for VKH is SO. That said, in the Masquerade Syndromes chapter of the Uveitis book, one condition is mentioned as masquerading for VKH. What is it?

**Number one on the DDx for VKH:** SO. **Number one on the DDx for SO:** VKH.

Sympathetic Ophthalmia (SO): Wadda ya know?
Number one on the DDx for VKH: **SO**.
Number one on the DDx for SO: **VKH**.

Sympathetic Ophthalmia (SO): Wadda ya know?

Recall this slide from earlier in the set. As stated then, tops on the DDx for VKH is SO. That said, in the Masquerade Syndromes chapter of the Uveitis book, one condition is mentioned as masquerading for VKH. What is it? Bilateral diffuse uveal melanocytic proliferation (BDUMP)
In a word, what sort of condition is BDUMP?
A paraneoplastic syndrome

Wadda ya know?

Recall this slide from earlier in the set. As stated then, tops on the DDx for VKH is SO. That said, in the Masquerade Syndromes chapter of the Uveitis book, one condition is mentioned as masquerading for VKH. What is it?
Bilateral diffuse uveal melanocytic proliferation (BDUMP)

In a word, what sort of condition is BDUMP?
A paraneoplastic syndrome

With which malignancies is it associated?
Gynecologic in women; lung and pancreatic in men

What is the classic presenting ocular complaint?
Bilateral rapid vision loss

What classic DFE finding puts it on the DDx for VKH?
Bilateral serous/exudative RDs

What are the two other classic findings on exam?
Bilateral…
--rapid cataract progression
--multiple large 'nevi' of the posterior choroid

What is the prognosis?
Poor, in terms of both vision and life expectancy
In a word, what sort of condition is BDUMP?
A paraneoplastic syndrome

Wadda ya know?

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Wadda ya know?
In a word, what sort of condition is BDUMP?
A paraneoplastic syndrome

Is BDUMP common, or rare?

Recall this slide from earlier in the set. As stated then, tops on the DDx for VKH is SO. That said, in the Masquerade Syndromes chapter of the Uveitis book, one condition is mentioned as masquerading for VKH. What is it?
Bilateral diffuse uveal melanocytic proliferation (BDUMP)

In a word, what sort of condition is BDUMP?
A paraneoplastic syndrome

Is BDUMP common, or rare?

Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associated?
Gynecologic in women; lung and pancreatic in men

What is the classic presenting ocular complaint?
Bilateral rapid vision loss

What classic DFE finding puts it on the DDx for VKH?
Bilateral serous/exudative RDs

What are the two other classic findings on exam?
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Is BDUMP common, or rare?
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Who is the typical BDUMP pt?
In a word, what sort of condition is BDUMP?
A paraneoplastic syndrome

Is BDUMP common, or rare?
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Is there a gender predilection?
In a word, what sort of condition is BDUMP?
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No

What is the classic presenting ocular complaint?
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What are the two other classic findings on exam?
Bilateral…
--rapid cataract progression
--multiple large 'nevi' of the posterior choroid

What is the prognosis?
Poor, in terms of both vision and life expectancy
Number one on the DDx for VKH: SO.
Number one on the DDx for SO: VKH.

So let's compare and contrast them!

Recall this slide from earlier in the set. As stated then, tops on the DDx for VKH is SO. That said, in the Masquerade Syndromes chapter of the Uveitis book, one condition is mentioned as masquerading for VKH. What is it? Bilateral diffuse uveal melanocytic proliferation (BDUMP).

In a word, what sort of condition is BDUMP? A paraneoplastic syndrome.

Is BDUMP common, or rare? Quite rare (as in, <50 or so reported cases).

Who is the typical BDUMP pt? An adult 50+.

Is there a gender predilection? No.

With which malignancies is it associated?

Is BDUMP common, or rare? Quite rare (as in, <50 or so reported cases).

Who is the typical BDUMP pt? An adult 50+.

Is there a gender predilection? No.

With which malignancies is it associated?

Gynecologic in women; lung and pancreatic in men.

What is the classic presenting ocular complaint? Bilateral rapid vision loss.

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--rapid cataract progression
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Q
Number one on the DDx for VKH: SO.
Number one on the DDx for SO: VKH.

So let's compare and contrast them!

Recall this slide from earlier in the set. As stated then, tops on the DDx for VKH is SO. That said, in the Masquerade Syndromes chapter of the Uveitis book, one condition is mentioned as masquerading for VKH. What is it?

Bilateral diffuse uveal melanocytic proliferation (BDUMP)

In a word, what sort of condition is BDUMP?
A  paraneoplastic syndrome

Is BDUMP common, or rare?
Quite rare (as in, <50 or so reported cases)

Who is the typical BDUMP pt?
An adult 50+

Is there a gender predilection?
No

With which malignancies is it associated?
Gynecologic in women; lung and pancreatic in men

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Bilateral rapid vision loss

What classic DFE finding puts it on the DDx for VKH?
Bilateral serous/exudative RDs

What are the two other classic findings on exam?
Bilateral…
--rapid cataract progression
--multiple large 'nevi' of the posterior choroid

What is the prognosis?
Poor, in terms of both vision and life expectancy
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Vogt-Koyanagi-Harada (VKH) Syndrome: Wadda ya know?
Sympathetic Ophthalmia (SO): Wadda ya know?

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