- 1. There should be no history of ocular trauma or surgery
- 2. There should be no clinical or laboratory evidence of other ocular disease entities
- 3. The uveitis must be bilateral exhibiting either A or B
 - A) Early Disease
 - i) Diffuse Choroiditis with either focal or bullous subretinal fluid
 - ii) If fundal appearances are equivocal must have:
 - a) Focal Choroidal perfusion delay, pinpoint leakage, placoid fluorescence and optic nerve staining on fluorescein angiography and
 - b) Diffuse choroidal thickening but no scleritis
 - B) Late Disease
 - i) Suggestive history of 3A and either ii or iii
 - ii) Depigmentation, either sunset glow or Sugiura's sign
 - iii) Nummular chorioretinal scars with retinal pigment epithelium (RPE) clumping and migration and recurrent chronic anterior uveitis
- 4. Active or history of either meningism, tinnitus, cerebrospinal fluid pleocytosis
- 5. Alopecia, Poliosis, or vitiligo

Complete VKH Syndrome requires Criteria 1-5, all inclusive

Incomplete VKH Syndrome requires Criteria 1-3 and either 4 or 5.

Probable VKH requires Criteria 1-3

Figure 1: Revised International Diagnostic Criteria for Vogt-Koyanagi-Harada (VKH)

Corticosteroids
Oral prednisone 100-200 mg initially, followed by gradual taper over 3-6 months
Pulse dose of methylprednisolone 1g/day for 3 days, followed by gradual tapering of oral prednisone over 3-6 months
Intravenous methylprednisolone 100-200 mg/day for 3 days followed by gradual tapering of oral prednisone over 3-6 months
Cyclosporin 5 mg/kg per day
Azathioprine 1-2.5 mg/kg per day
Biologics
Anti-TNF-α monoclonal antibody

Figure 2: Treatment options in VKH [1]

Subjective:

- Previously healthy 34 year old Metis female
- Three week history of pain, redness, blurred vision and increased sensitivity to light in both eyes, previously diagnosed as bilateral uveitis.
- Prior to eye complaints, experienced chills and headache with acute illness. Significant family history of ankylosing spondylitis.
- Recent development of mild ringing of the ears, bilateral, and patches of pale skin appearing in her inguinal area on right side.

Objective:

- Ophthalmological exam: best corrected visual acuity 20/40 OD and 20/30 OS with intra ocular pressures of 11 and 12, respectively.
- Dilated fundoscopic exam: Anterior chamber revealed occasional cell and 2+ cells, left and right respectively. Posterior chamber revealed + cells, bilaterally.
- Significant optic nerve edema 360 degrees with serous retinal detachments in right eye. Significant macular edema with a serous retinal detachment in left eye.
- Fluoroscein Angiogram: diffuse choroiditis with focal delays in choroidal perfusion and multifocal areas of pinpoint leakage, bilaterally
- CBC, Lipids ANA, glucose, Lyme serology, syphilis, serum ACE, ESR, CRP, serum protein electrophoresis, serology for cat scratch and 120 degree Visual Field test were unremarkable.
- CSF revealed pleocytosis but culture was negative for bacteria.

Assessment:

- VKH, as patient met diagnostic criteria outlined by the American Uveitis Society
- Bilateral anterior uveitis.

Plan:

- Continue treatment for uveitis which includes diclofenac (Voltaren) drops, prednisolone (Pred Forte) drops and olfloxacin (Ocuflox) drops, bilaterally. Cyclopentolate (Cyclogyl) drops for right eye only.
- Systemic steroid therapy to treat VKH using IV methylprednisolone 1 gram x 3 days and then oral prednisone 80 mg daily on a tapering dose for 3-6 months