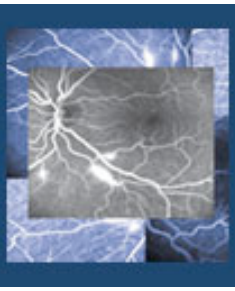


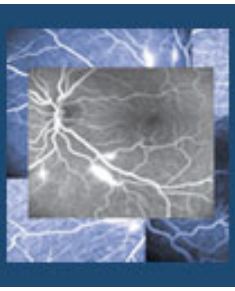
# Susac's Syndrome

**Symptoms and Other Abnormalities Associated with Susac's Syndrome**
Because Susac's Syndrome is due to injury to the microvasculature in the brain, retina, and inner ear, it causes the following categories of symptoms:

- Brain Symptoms----Cognitive dysfunction and other neurologic symptoms
- Eye Symptoms----Visual disturbance
- Inner Ear Symptoms----Hearing loss, Vertigo, Tinnitus

**Brain Symptoms can include:**

- Headache
  - often with vomiting,
  - often migraine-like
- Memory loss
- Confusion
- Disorientation
- Disorganized thought
- Poor awareness of surroundings
- Compromised insight into one's own problems/illness
- Slowed thought processing
- Impaired reasoning
- Impaired judgment
- Impaired executive function
- Less alert, mentally
- Lethargy
- Apathy, vagueness
- Drowsiness, excessive sleeping
- Obtundation/Unresponsiveness (rarely)
- Difficulty with focusing, concentrating, maintaining attention
- Difficulty with arithmetic
- Difficulty finding, recognizing, or speaking words
- Decreased verbal fluency
- Incoherent speech
- Dysarthria (slurred speech)
- Personality change
- Compromised ability to express emotions
- Emotional lability (widely swinging emotions)
- Psychiatric disturbance
  - Fluctuating depression
  - Mood disturbance
  - Agitation
  - Severe anxiety
  - Aggressive behavior
  - Confabulation
  - Acute psychosis
  - Delusions, hallucinations, paranoia
  - Withdrawn, irritable
  - Disinhibition, inappropriate gestures
  - Perseveration
  - Poor cooperation
- Loss of balance (ataxia)
- Weakness in arm, leg (motor hemiparesis)
- Numbness in arm, leg, face, tongue, lip (sensory hemiparesis)
- Incontinence, neurogenic bladder
- Pseudobulbar affect and speech
- Frontal lobe syndrome
- Seizures



# Susac's Syndrome

## Eye Symptoms

- "Dark spot," "black spot," "pink spot," or "shadow" in one part of visual field, such that the patient "can't see anything" in that spot. (This is called a "scotoma.") Other less specific symptoms due to the same thing might be expressed as:
  - Visual blurring
  - Obscured vision
  - Visual disturbance
  - "Flashing lights"
- Loss of peripheral vision (constriction of visual fields)
- Diplopia (double vision)---rare
- Inverted vision---very rare

## Inner Ear Symptoms

- Hearing loss"
- Vertigo ("whirling" dizziness)
- Tinnitus (ringing in the ears)

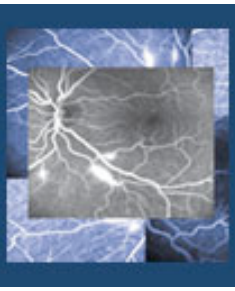
## Neurologic Abnormalities upon Physical Examination

The neurologic exam is often normal. But, the following can occur:

- Ataxia (loss of balance)
- Impaired tandem gait
- Motor hemiparesis
- Sensory hemiparesis
- Hyperactive tendon reflexes
- Spasticity
- Extensor plantar responses (+ Babinski)
- Dysmetria
- Dysdiadochokinesia
- Apraxia
- Frontal lobe findings---grasp reflex
- Pseudobulbar affect
- Sphincter deficit

## Ophthalmologic Abnormalities

- Fundoscopic abnormalities:
  - "Cotton wool" spots
  - Arterial narrowing
  - Perivascular sheathing
  - "Silver streaks"
  - "Pearls," "string of pearls"
  - Gass plaques
  - Residual ischemic retinal changes
  - Optic disc pallor
- Visual field testing:
  - Visual field deficit
- OCT (Ocular Coherence Tomography):
  - Thinning of the nerve fiber layer
- Fluorescein Angiography abnormalities:
  - Branch retinal artery occlusion
  - "Leakage" and vessel wall hyperfluoresence ("staining")
  - Peripheral shunting/collateral formation/artery-to-artery anastomosis
  - Capillary "drop-out."
  - Neovascularization
- Visual acuity is usually normal.
- Other:
  - Nystagmus
  - Gaze palsy



# Susac's Syndrome

## MRI Abnormalities

- Abnormalities in the Corpus Callosum:
  - "Snowballs"
  - Linear defects ("Spokes")
  - "Holes"

  These corpus callosal abnormalities are primarily located in the central portion of the corpus callosum, as opposed to the under-surface of the corpus callosum.

  Callosal atrophy commonly occurs.
- Other white matter lesions, in the following locations:
  - Periventricular
  - Centrum semiovale
  - Subcortical white matter
  - Cerebellum
  - Middle Cerebellar peduncles
  - Brain stem
- Gray matter lesions:
  - Cortex
  - Deep Gray matter lesions
    - Thalamus
    - Basal ganglia
- Leptomeningeal enhancement

## CSF Findings

- Elevated CSF protein
- Pleocytosis (elevated WBC)---mild
- Oligoclonal bands are usually absent; rarely present
- Myelin basic protein level can be elevated

## Audiological Evaluation

- Sensorineural hearing loss:
  - Typically at low frequencies, or low-mid frequencies
  - Sometimes at all frequencies
  - Sometimes at low and high frequencies
  - Rarely at high frequencies only
- Poor speech discrimination