


Visual Snow Syndrome



Normal vision | Visual snow

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Financial Disclosures

- No relevant disclosures

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Case presentation

- 14 yo WF
- Cc: “pixelated” vision
 - TV static in her entire visual field
 - 24/7
 - Afterimages
 - Photophobia
- PMH
 - Migraine headaches

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Case

- Va 20/20 OU
- Pupils normal, no APD
- Ext'l/SLE normal
- Motility normal, ortho
- HVF normal
- IOP normal
- Fundi normal

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Case

- Cc: "Pixelated" vision
- Dx?
- Work-up?
- Treatment?



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DDX: Spots & Dots

- Diagnosis – benign vs not-so-benign
 - Benign
 - Vivid imagination
 - Normal entoptic phenomenon
 - Vitreous floaters
 - Purkinje tree
 - Blue field phenomenon

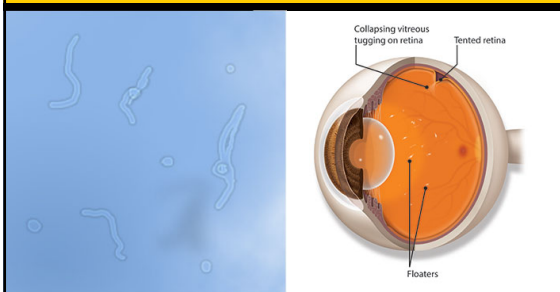
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Entoptic phenomenon

- Visualize an internal structure in the eye
- Examples
 - Floaters
 - Vitreous strands
 - Hemorrhage

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Entoptic Phenomenon: Floaters

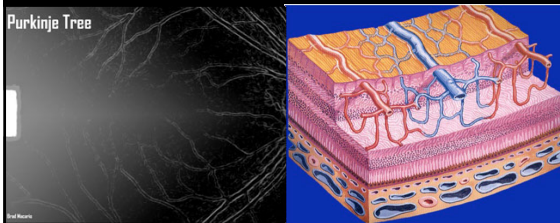


Boldedscience.com

Asa.org

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Entoptic Phenomenon: Purkinje Tree



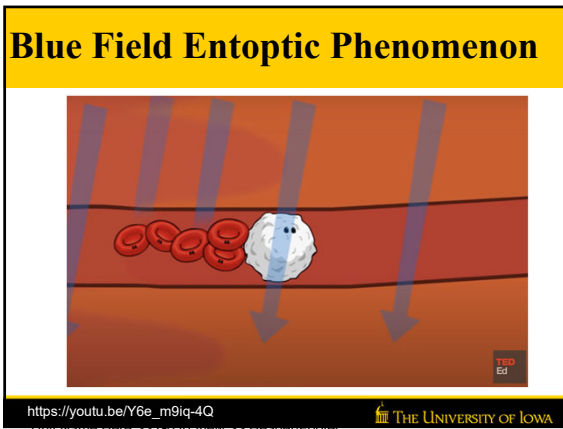
isa More

http://en.wikipedia.org/wiki/File:Purkinje_Tree_BM.jpg

Retinadoctor.com.au

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Entoptic Phenomenon: Phosphenes

- Phosphenes
 - Perception of light w/o light stimulus
 - Mechanical stimulation of the retina
 - Eye rubbing
 - Valsalva

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Entoptic Phenomenon: Phosphenes



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DDX: Spots & Dots

- Diagnosis – not-so-benign
 - Migraine
 - Occipital lobe epilepsy/lesion
 - Paraneoplastic retinopathy
 - White dot syndromes

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Migraine Visual Aura



Migraine Visual Aura



Occipital lobe epilepsy/lesion

OCCIPITAL LOBE

The occipital lobe has its name because it rests below the occipital bone of the skull. It is also the smallest of the lobes. In general, the occipital lobe deals with aspects of vision. Seizures can spread and cause focal impaired awareness seizures or focal motor seizures.

IT PLAYS A KEY ROLE IN

Visual Distance	Colour Determination	Object Recognition	Face Recognition	Depth Perception
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SOME COMMON SEIZURE SYMPTOMS

Visual Phenomena e.g. coloured flashes or flashes	Visual Loss e.g. loss part of visual field or blindness	Visual Hallucinations e.g. animals, people or scenes	Repetitive Uncontrolled Eye Movements
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Neuro-o.se

Facebook.com

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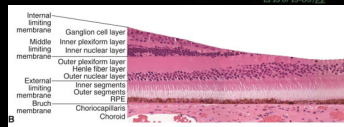
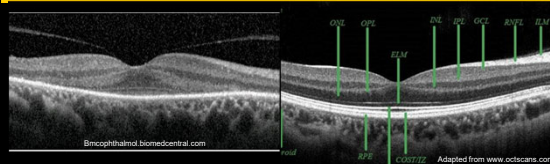
Paraneoplastic/AI retinopathy



<https://www.youtube.com/watch?v=902CP16u910>

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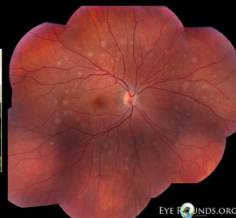
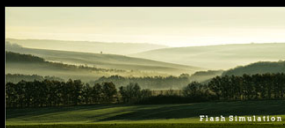
Paraneoplastic retinopathy: e.g., cancer associated retinopathy



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White Dot Syndromes e.g., multiple evanescent white dot syndrome

- Photopsias



EYE FUNDUS.ORG

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Visual Snow: Aeropsia

- TV static
- Constant 24/7
- Flickering dots/lights
- Superimposed on environment



Twitter.com

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Visual Snow: Who Knows About It?

Visual Snow



Visual Snow



Visual Snow

- <http://www.visionsimulations.com/>

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Symptoms of Visual Snow

- A million dots
- Black cracks and lines
- Blobs of white and gray
- Blue squares
- Carpet background
- Circles
- Clouds
- Comets
- Dots, b/w & colored
- Grainy vision
- Heat waves
- Lights, flash/flicker
- Lines of ants
- Photopsias
- Rain-like pattern
- Snow
- Squiggles
- TV static

Liu et al. Neurology 45:664-668, 1995



Visual Snow: many names

- Prolonged migraine aura status (Haas 1982)
- Sustained visual aura
- Persistent positive visual phenomena (1995)
- Persistent migraine aura
- Migraine aura status
- Persistent migrainous visual phenomena
- Persistent migraine aura w/o infarction (IHC 2003)
- Primary persistent visual disturbance (2005)
- Visual snow (2014)



Persistent Aura w/o Infarction

- Persistent aura without infarction
 - Aura symptoms > 1 week w/o infarction
 - Rare
 - Bilateral
 - Lasts months to years
 - Most have h/o migraine or variant



Primary Persistent Visual Disturbance

doi:10.1111/j.1468-2982.2004.00858.x

Diffusion- and perfusion-weighted MR imaging in persistent
migrainous visual disturbances

HR Jäger¹, NJ Giffin & PJ Goadsby
Headache Group and Division of Neuroradiology, Institute of Neurology, The National Hospital for Neurology and Neurosurgery, London, UK

Cephalalgia

Jäger HR, Giffin NJ & Goadsby PJ. Diffusion- and perfusion-weighted MR imaging in persistent migrainous visual disturbances. *Cephalalgia* 2005; 25:323-332. London. ISSN 0333-1024

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Visual Snow

doi:10.1093/brain/awu050

Brain 2014; 137: 1419-1428 | 1419

BRAIN
A JOURNAL OF NEUROLOGY

**'Visual snow' – a disorder distinct from persistent
migraine aura**

Christoph J. Schankin,^{1,2,*} Farooq H. Maniyar,^{1,2} Kathleen B. Digre³ and Peter J. Goadsby^{1,2}

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Visual Snow Syndrome: Criteria

- Dynamic, continuous, flickering tiny dots
- Entire visual field
- 2 of the following 4
 - Enhanced entoptic phenomenon
 - Photophobia
 - Nyctalopia
 - Palinopsia

Curr Opin Neurol 2018;31:52-58

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VSS: Criteria

- Entoptic phenomenon
 - Floaters 81%
 - Blue field entoptic 79%
- Photophobia 74%
- Nyctalopia 68%

Boldscience.com

JNO 38(4):514-521, 2018

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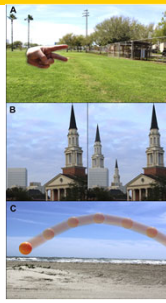
Palinopsia

- Perseveration of visual image
 - Stimulus gone
 - Different than normal retinal afterimage

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VSS: Criteria

- Palinopsia 69%
- Stationary 86
- Trailing moving objects 60

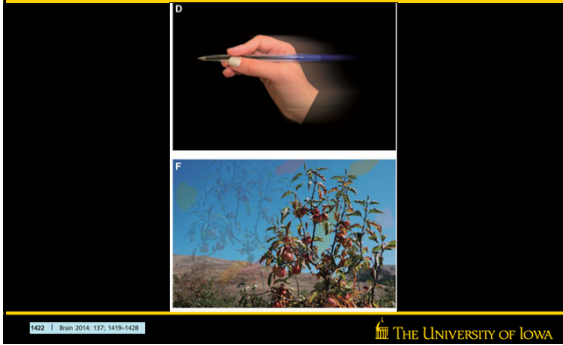


Surv Ophthalmol 60:1-35, 2015

JNO 38(4):514-521, 2018

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Palinopsia



VSS: Criteria

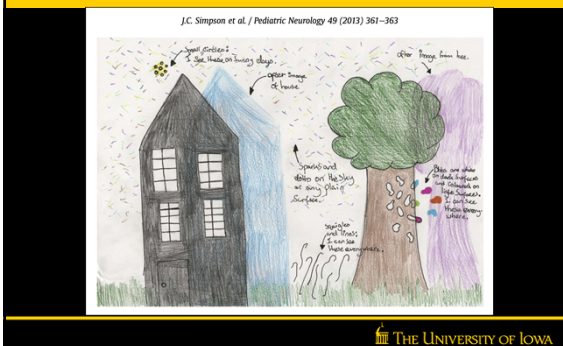
- Other frequent & persistent visual phenomenon
 - Halos
 - Metamorphopsia
 - Colored clouds
 - Water running down a window



Curr Neurol Neurosci Rep 2018;18:52
Brain 2014;137:1419-1428

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VSS



Visual Snow



VSS: Demographics

- Web-based survey of 1,104 pts w/VS
- Xage 29
- M=F
- Onset early in life
- All of their life 40%

Neurology 2020;94:e564-574. doi:10.1212 THE UNIVERSITY OF IOWA

VSS: Comorbidities

- Comorbidities
 - Migraine 60%
 - Often severe migraine triggers
 - Tinnitus 62%
 - Concentration difficulties 60%
 - Anxiety 44%
 - Depression 19%

JNO 38(4):514-521, 2018 THE UNIVERSITY OF IOWA

VSS: Testing normal

- Ophthalmologic examination
- Neurologic examination
- Ancillary testing VF
 - OCT
 - ERG
 - VEP
- MRI

VSS: Diagnosis

- Mistakenly considered psychogenic
- Symptoms too stereotypical

VSS: Pathophysiology

- Unknown
- Bilateral, symmetric, constant, spacing
- Cortical
- Normal?
 - Visual noise?
 - Similar to tinnitus?
 - Result of abnormal inhibition?

VSS: Pathophysiology

- 3 major theories:
 - Hyperexcitability
 - Hypoconnectivity
 - Thalamocortical dysfunction
 - Combination of all three

VSS: Pathophysiology

- Cortical hyperexcitability
 - Magnetoencephalography
 - Functional MRI abnormalities
 - Hypermetabolism on PET
 - Elevated spontaneous neural firing
 - Inconsistent studies VEP and EEG

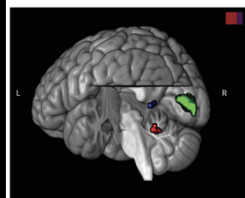


<https://brainpanter.csail.mit.edu/>

Neurology 2018; 88:1243-49 Brain. 2011;134:2387-2395
 Headache 2014;54:957-66 Headache 2015;55(10):1436-41

VSS: Pathophysiology

Figure 3 Summary of cortical volumetric changes in patients with visual snow syndrome, as shown by voxel-based morphometry



Render illustration of the 3 brain regions of increased gray matter volume in patients with visual snow syndrome, with respect to healthy controls. Left V1 cluster is illustrated in green; left V2 cluster in blue; left cerebellum cluster in red. For statistical values of each area, see the text. Image was created in MRICROGL and superimposed on standard brain template.

Neurology 95(13):e1792-31799, 2020

VSS: Pathophysiology

FrontNeuroL 2019; 10: 1325 THE UNIVERSITY OF IOWA

VSS: Treatment

•Review of the literature

Medication	Overall trials	Ineffective	Effective (total response)
Lamotrigine	36	28	8 (1)
Topiramate	13	11	2
Valproate	12	11	1 (1)
Propranolol	6	5	1
Sertraline	5	4	1
Verapamil	3	2	1
Naproxen	2	1	1
Baclofen	1	0	1

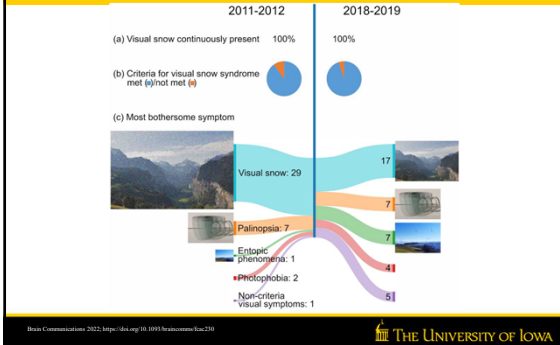
Prog Brain Res 2020;255:311-316 THE UNIVERSITY OF IOWA

VSS: Treatment

- Reassurance and validation
- Colored filters (blue 80% pts)
- Education
 - Visionsimulations.com
 - Visualsnowinitiative.org
 - Wikipedia.com
 - Eyeonvision.org

Neurology 2019;93:e3980e403
J Clin Neuroscience 2016; 28:123-127. THE UNIVERSITY OF IOWA

Prognosis



VSS: Over the years experience

- Nearly 100 patients over 33 years
- Patients wide age range, most 20-30 yrs
- Highly observant, articulate
- Symptoms stereotypical
- Examinations normal
- Treatments unsuccessful
- Tale of a neurologist

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VSS: Conclusions

- History is key
- Ophthalmologic examination normal



padersophia.wordpress.com

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VSS: Conclusions

- Criteria
 - Bilateral constant flickering tiny dots
 - 2 of the following 4
 - Palinopsia
 - Enhanced entoptic phenomenon
 - Photophobia
 - Nyctalopia

padersophia.Wordpress.com

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VSS: Conclusions

- | | | |
|------|-----------------------------------|--|
| •DDX | Benign | Not-so-benign |
| | Entoptic Phenomenon
Phosphenes | Migraine
Seizure
Paraneoplastic
retinopathies
White dot syndrome |
- Treatment
 - Reassurance
 - Education
 - Visualsexperiences.org
 - Visionsimulations.com

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